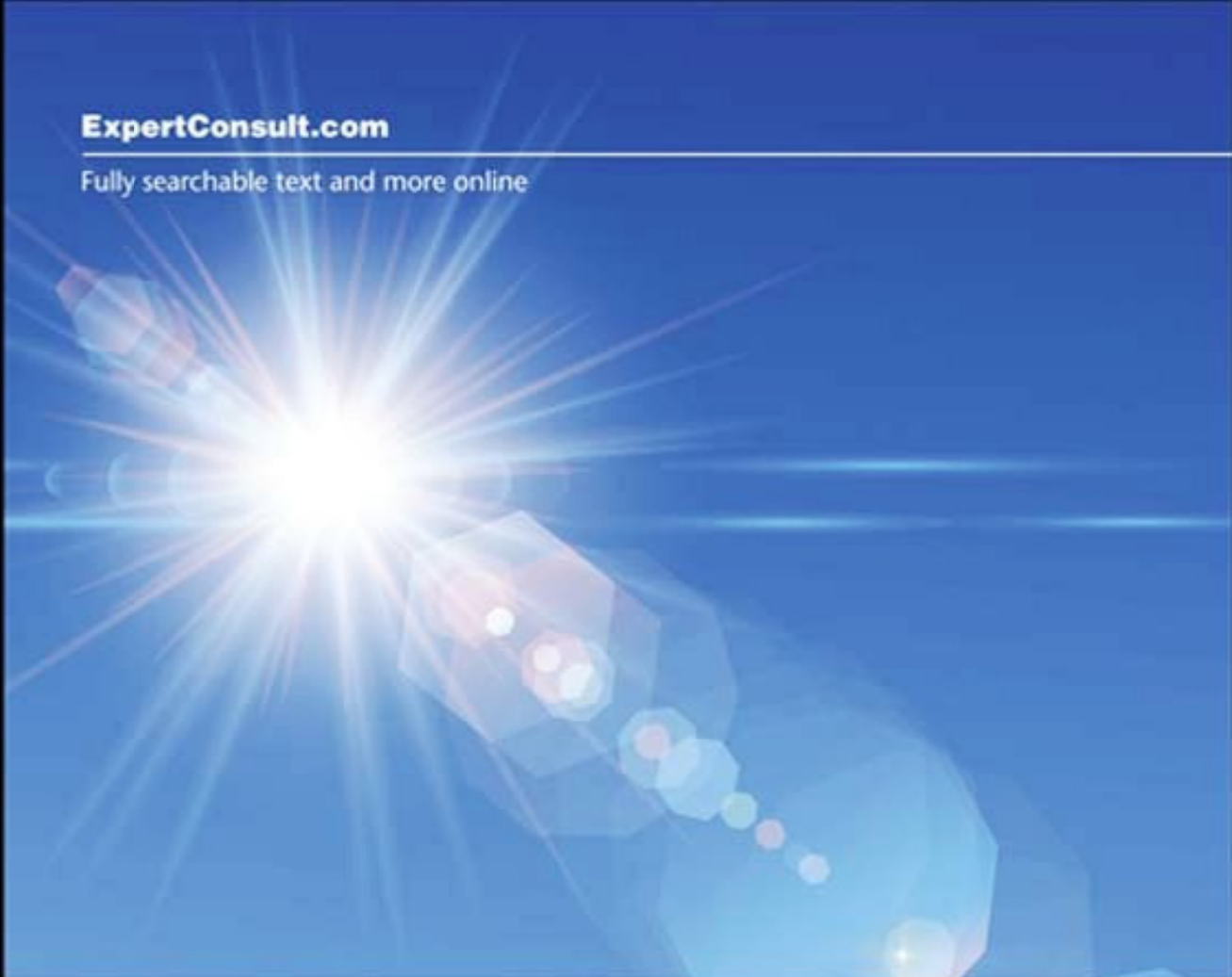


ExpertConsult.com

Fully searchable text and more online



Kanski's

Clinical Ophthalmology

A SYSTEMATIC APPROACH

EIGHTH EDITION

Brad Bowling

ELSEVIER



Kanski's Clinical Ophthalmology

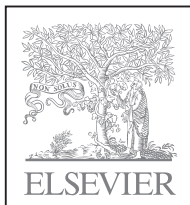
A SYSTEMATIC APPROACH

EIGHTH EDITION

Brad Bowling

FRCSEd(Ophth), FRCOphth, FRANZCO

Ophthalmologist
Sydney
New South Wales
Australia



For additional online content visit [expertconsult](http://expertconsult.com)

Chapter Orbit 3

INTRODUCTION 78

Anatomy 78
Clinical features 78
Investigation 81

THYROID EYE DISEASE 82

Introduction 82
Clinical features 82
Investigation 84
Treatment 84

INFECTIONS 87

Preseptal cellulitis 87
Bacterial orbital cellulitis 87
Rhino-orbital mucormycosis 89

NON-INFECTIVE INFLAMMATORY DISEASE 89

Idiopathic orbital inflammatory disease 89
Orbital myositis 90
Acute dacryoadenitis 91
Tolosa-Hunt syndrome 92
Wegener granulomatosis 92

NON-NEOPLASTIC VASCULAR ABNORMALITIES 92

Cavernous sinus thrombosis 92
Carotid-cavernous fistula 93

CYSTIC LESIONS 95

Dacryops 95
Dermoid cyst 95
Sinus mucocoele 96
Encephalocoele 97

VASCULAR TUMOURS 98

Varices 98
Lymphangioma 98
Capillary haemangioma 100
Cavernous haemangioma 102

LACRIMAL GLAND TUMOURS 103

Pleomorphic lacrimal gland adenoma 103
Lacrimal gland carcinoma 105

NEURAL TUMOURS 106

Optic nerve glioma 106
Optic nerve sheath meningioma 107
Plexiform neurofibroma 109
Isolated neurofibroma 109

LYMPHOMA 109

RHABDOMYOSARCOMA 109

METASTATIC TUMOURS 110

Adult metastatic tumours 110
Childhood metastatic tumours 111
Orbital invasion from adjacent structures 112

THE ANOPHTHALMIC SOCKET 113

Surgical procedures 113
Rehabilitation 114

CRANIOSYNOSTOSES 117

INTRODUCTION

Anatomy

The orbit is a pear-shaped cavity, the stalk of which is the optic canal (Fig. 3.1).

- **The roof** consists of two bones: the lesser wing of the sphenoid and the orbital plate of the frontal bone. It is located subjacent to the anterior cranial fossa and the frontal sinus. A defect in the orbital roof may cause pulsatile proptosis due to transmission of cerebrospinal fluid pulsation to the orbit.
- **The lateral wall** also consists of two bones: the greater wing of the sphenoid and the zygomatic. The anterior half of the globe is vulnerable to lateral trauma since it protrudes beyond the lateral orbital margin.
- **The floor** consists of three bones: the zygomatic, maxillary and palatine. The posteromedial portion of the maxillary bone is relatively weak and may be involved in a 'blowout' fracture (see Ch. 21). The orbital floor also forms the roof of the maxillary sinus so that maxillary carcinoma invading the orbit may displace the globe upwards.
- **The medial wall** consists of four bones: maxillary, lacrimal, ethmoid and sphenoid. The lamina papyracea, which forms part of the medial wall, is paper-thin and perforated by numerous foramina for nerves and blood vessels. Orbital cellulitis is therefore frequently secondary to ethmoidal sinusitis.
- **The superior orbital fissure** is a slit linking the cranium and the orbit, between the greater and lesser wings of the sphenoid bone; through it pass numerous important structures.
 - The superior portion contains the lacrimal, frontal and trochlear nerves, and the superior ophthalmic vein.
 - The inferior portion contains the superior and inferior divisions of the oculomotor nerve, the abducens and nasociliary nerves, and sympathetic fibres from the cavernous plexus.
 - Inflammation of the superior orbital fissure and apex (Tolosa–Hunt syndrome) may therefore result in a multitude of signs including ophthalmoplegia and venous outflow obstruction.
- **The inferior orbital fissure** lies between the greater wing of the sphenoid and the maxilla, connecting the orbit to the pterygopalatine and infratemporal fossae. Through it run the maxillary nerve, the zygomatic nerve and branches of the pterygopalatine ganglion, as well as the inferior ophthalmic vein.

Clinical features

Symptoms

Symptoms of orbital disease include eyelid and conjunctival swelling, redness, watering, pain (sometimes on, or exacerbated by, eye movement), increasing ocular prominence, displacement or a sunken impression of the eye, double vision and blurring, and sometimes a pulsing sensation or audible bruit.

Soft tissue involvement

Eyelid and periocular oedema, skin discoloration, ptosis, chemosis (oedema of the conjunctiva, which may involve the plica and caruncle) and epibulbar injection (Fig. 3.2) may be seen; causes include thyroid eye disease, orbital inflammatory diseases and obstruction to venous drainage.

Proptosis

Proptosis (Fig. 3.3) describes an abnormal protrusion of an organ, but is generally applied to the eyeball; exophthalmos refers

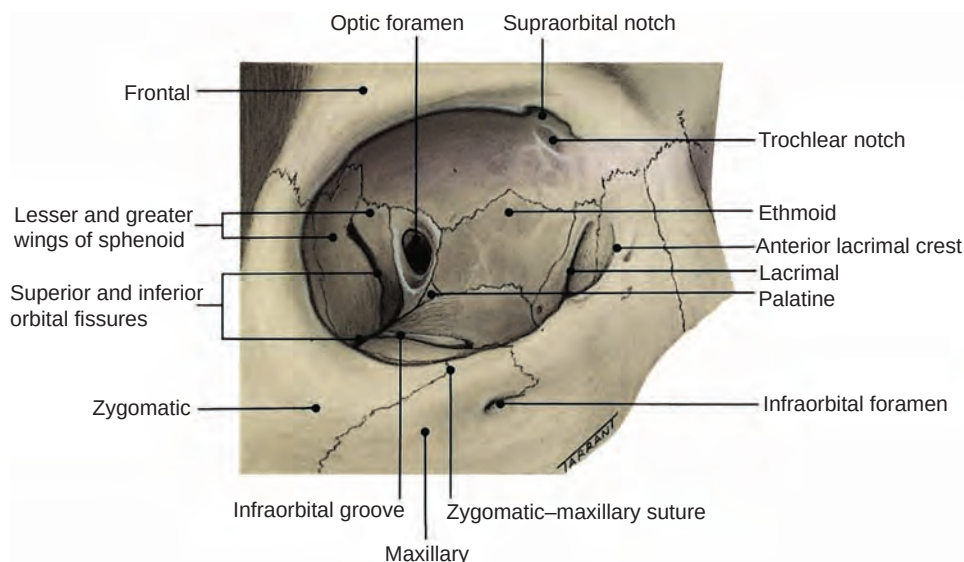


Fig. 3.1 Anatomy of the orbit

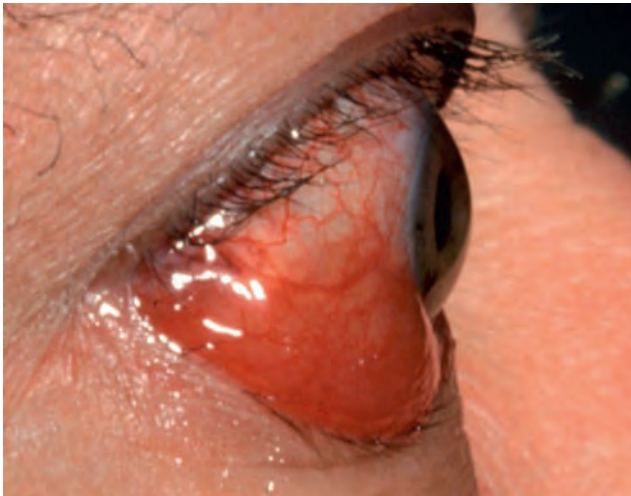


Fig. 3.2 Chemosis and injection in orbital disease

specifically to the eyeball only. Proptosis may be caused by retrobulbar lesions or, less frequently, a shallow orbit. The intraorbital portion of the optic nerve is longer (25 mm) than the distance between the back of the globe and the optic canal (18 mm). This allows for significant forward displacement of the globe (proptosis) without excessive stretching of the nerve.

- **Asymmetrical proptosis** is readily detected by looking down at the patient from above and behind (Fig. 3.4A).
- **The direction** of proptosis may indicate the likely pathology. For example, space-occupying lesions within the muscle cone such as a cavernous haemangioma or optic nerve tumours cause axial proptosis, whereas extraconal lesions usually give rise to combined proptosis and dystopia (see next).
- **Dystopia** implies displacement of the globe in the coronal plane, usually due to an extraconal orbital mass such as a lacrimal gland tumour (Fig. 3.4B). Horizontal displacement is measured from the midline (nose) to the centre of the pupil while vertical dystopia is read on a vertical scale perpendicular to a horizontal rule placed over the bridge of the nose. The measured eye should fixate straight ahead, if necessary facilitating this by occluding the fellow eye.
- **The severity** of proptosis can be measured with a plastic rule resting on the lateral orbital margin, or with the Luedde™ exophthalmometer using a similar principle. Commonly, a binocular exophthalmometer (e.g. Hertel) is employed, using visualization of the corneal apices to determine the

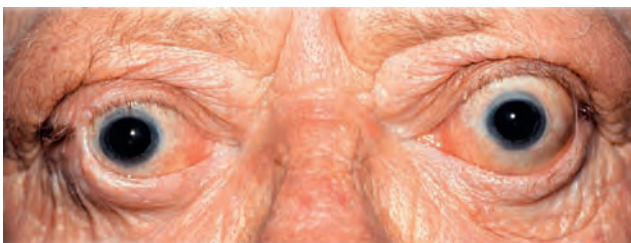


Fig. 3.3 Bilateral proptosis
(Courtesy of C Barry)



Fig. 3.4 General signs of orbital disease. (A) Left proptosis visualized from above; (B) right inferior dystopia; (C) measurement of proptosis with an exophthalmometer

degree of ocular protrusion from a scale (Fig. 3.4C). Measurements can be taken both relaxed and with the Valsalva manoeuvre. Readings greater than 20 mm are indicative of proptosis and a difference of 2–3 mm or more between the two eyes is suspicious regardless of the absolute values. The dimensions of the palpebral apertures and any lagophthalmos should also be noted.

- **Pseudoproptosis** (the false impression of proptosis) may be due to facial asymmetry, enlargement of the globe (e.g. high myopia or buphthalmos), lid retraction or contralateral enophthalmos.

Enophthalmos

Enophthalmos implies recession of the globe within the orbit. Causes include congenital and traumatic orbital wall abnormalities, atrophy of the orbital contents (e.g. radiotherapy, scleroderma, chronic eye poking in blind infants – the ‘oculodigital’ sign) or sclerosis (e.g. metastatic scirrhous carcinoma, sclerosing orbital inflammatory disease). Pseudoenophthalmos may be caused by a small or shrunken eye (microphthalmos or phthisis bulbi), by ptosis, or by contralateral proptosis or pseudoproptosis.

Ophthalmoplegia

Defective ocular motility is very common in orbital disease. Causes include an orbital mass, restrictive myopathy (e.g. thyroid eye disease – Fig. 3.5, orbital myositis, tethering of muscles or tissue after orbital wall fracture), ocular motor nerve involvement associated with lesions in the cavernous sinus, orbital fissures or posterior orbit (e.g. carotid–cavernous fistula, Tolosa–Hunt syndrome, malignant lacrimal gland tumours). The following tests may be used to differentiate a restrictive from a neurological motility defect:

- **Forced duction test.** Under topical anaesthesia, the insertion of the muscle in an involved eye is grasped with forceps and the globe rotated in the direction of reduced mobility; checked movement of the globe indicates a restrictive problem; no resistance will be encountered with a neurological lesion.
- **Differential intraocular pressure (IOP) test** involves less discomfort than forced duction and an objective rather than

subjective endpoint. The IOP is measured in the primary position of gaze and then with the patient attempting to look in the direction of limited mobility; an increase of 6 mmHg or more denotes resistance transmitted to the globe by muscle restriction (the Braley sign).

- **Saccadic eye movements** in neurological lesions are reduced in velocity, while restrictive defects manifest normal saccadic velocity with sudden halting of ocular movement.

Dynamic properties

- **Increasing venous pressure** by dependent head position, the Valsalva manoeuvre or jugular compression may induce or exacerbate proptosis in patients with orbital venous anomalies or infants with orbital capillary haemangioma.
- **Pulsation** is caused either by an arteriovenous communication or a defect in the orbital roof. In the former, pulsation may be associated with a bruit depending on the size of the communication. In the latter the pulsation is transmitted from the brain by the cerebrospinal fluid and there is no associated bruit. Mild pulsation is best detected on the slit lamp, particularly by applanation tonometry.
- **A bruit** is a sign found with a larger carotid–cavernous fistula. It is best heard with the bell of the stethoscope and is lessened or abolished by gently compressing the ipsilateral carotid artery in the neck.

Fundus changes

- **Optic disc swelling** may be the initial feature of compressive optic neuropathy (Fig. 3.6A).
- **Optic atrophy** (Fig. 3.6B), which may be preceded by swelling, is a feature of severe compressive optic neuropathy. Important causes include thyroid eye disease and optic nerve tumours.



Fig. 3.5 Restrictive myopathy and bilateral lid retraction and proptosis in thyroid eye disease – nine positions of gaze (Courtesy of C Barry)

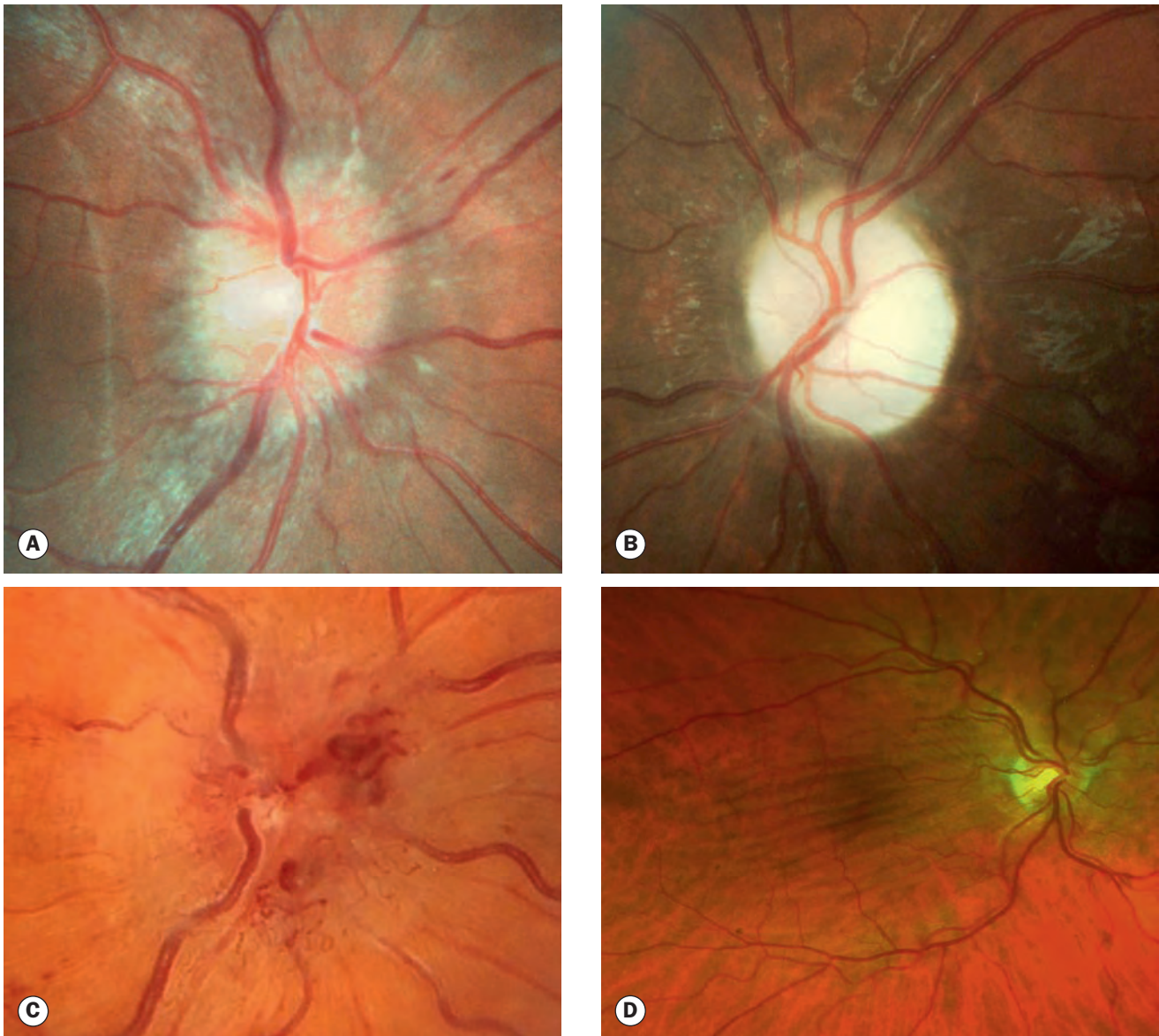


Fig. 3.6 Fundus changes in orbital disease. (A) Disc swelling; (B) optic atrophy; (C) opticociliary vessels on a chronically swollen disc; (D) choroidal folds

(Courtesy of S Chen – fig. D)

- **Opticociliary collaterals** consist of enlarged pre-existing peripapillary capillaries that divert blood from the central retinal venous circulation to the peripapillary choroidal circulation when there is obstruction of the normal drainage channels. On ophthalmoscopy the vessels appear as large tortuous channels most frequently sited temporally, which disappear at the disc margin (Fig. 3.6C). The collaterals may be associated with any orbital or optic nerve tumour that compresses the intraorbital optic nerve and impairs blood flow through the central retinal vein. The most common tumour associated with shunts is an optic nerve sheath meningioma but they may also occur with optic nerve glioma, central retinal vein occlusion, idiopathic intracranial hypertension and glaucoma.
- **Choroidal folds** (Fig. 3.6D) are discussed in detail in Ch. 14; they may occur in a wide variety of orbital lesions. Although tending to be more common with greater amounts of proptosis and anteriorly located tumours, in some cases their presence can precede the onset of proptosis.

Investigation

- **Computed tomography (CT)** is useful for depicting bony structures and the location and size of space-occupying lesions. It is of particular value in patients with orbital trauma because it can detect small fractures, foreign bodies, blood, herniation of extraocular muscle and emphysema (see Ch. 21). It is, however, unable to distinguish different

pathological soft tissue masses that are radiologically isodense. Confirmation of an orbital abscess in cellulitis is a relatively common indication.

- **Magnetic resonance imaging (MRI)** can demonstrate orbital apex lesions and intracranial extension of orbital tumours, and is useful for imaging orbital inflammatory disease. Serial short T₁ inversion recovery (STIR) scans are valuable in assessing inflammatory activity in thyroid eye disease (see Ch. 19).
- **Plain X-rays** are little used except for the initial diagnosis of traumatic bony injury.
- **Ultrasonography** can provide useful information, particularly with high-grade apparatus and an experienced operator, but does not image the orbital apex well.
- **Fine needle biopsy** is sometimes performed, particularly in suspected neoplastic disease. Potential problems include haemorrhage and ocular penetration.

THYROID EYE DISEASE

Introduction

Thyroid eye disease (TED), also known as thyroid-associated orbitopathy and Graves ophthalmopathy, is a very common orbital disorder, and is the most common cause of both bilateral and unilateral proptosis in an adult.

Thyrotoxicosis

Thyrotoxicosis (hyperthyroidism) is a condition involving excessive secretion of thyroid hormones. Graves disease, the most common form of hyperthyroidism, is an autoimmune disorder in which IgG antibodies bind to thyroid stimulating hormone (TSH) receptors in the thyroid gland and stimulate secretion of thyroid hormones. It is more common in females and may be associated with other autoimmune disorders. Presentation is often in the fourth or fifth decades with symptoms including weight loss despite good appetite, increased bowel frequency, sweating, heat intolerance, nervousness, irritability, palpitations, weakness and fatigue. There may be enlargement of the thyroid gland, tremor, palmar erythema, and warm and sweaty skin. Thyroid acropachy is a phenomenon similar to clubbing of the fingers, occurring in 1%; pretibial myxoedema (1–5%) is indurated thickening of the skin of the shins. Cardiac manifestations may include sinus tachycardia and other arrhythmias. Other autoimmune disorders can be associated. Thyroid function is commonly tested initially with a TSH level; if this is low, or normal but thyroid disease is still suspected, a range of additional investigations can be carried out. Treatment options include carbimazole, propylthiouracil, propranolol, thyroid ablation with radioactive iodine, and partial thyroidectomy.

Risk factors for ophthalmopathy

Once a patient has Graves disease, the major clinical risk factor for developing TED is smoking. The greater the number of cigarettes

smoked per day, the greater the risk, and giving up smoking seems to reduce the risk. Women are five times more likely to be affected by TED than men, but this largely reflects the increased incidence of Graves disease in women. Radioactive iodine used to treat hyperthyroidism can worsen TED. TED can also, though less commonly, occur in euthyroid and hypothyroid (including treated hyperthyroid) patients. It can sometimes be the presenting manifestation of thyroid-related disease.

Pathogenesis of ophthalmopathy

Thyroid ophthalmopathy involves an organ-specific autoimmune reaction in which an antibody that reacts against thyroid gland cells and orbital fibroblasts leads to inflammation of extraocular muscles, interstitial tissues, orbital fat and lacrimal glands characterized by pleomorphic cellular infiltration, associated with increased secretion of glycosaminoglycans and osmotic imbibition of water. There is an increase in the volume of the orbital contents, particularly the muscles, which can swell to eight times their normal size. There may be a secondary elevation of intraorbital pressure, and the optic nerve may be compressed. Subsequent degeneration of muscle fibres eventually leads to fibrosis, which exerts a tethering effect on the involved muscle, resulting in restrictive myopathy and diplopia.

Clinical features

Introduction

TED typically proceeds through a congestive (inflammatory) stage in which the eyes are red and painful; this tends to remit within 1–3 years and only about 10% of patients develop serious long-term ocular problems. A fibrotic (quiescent) stage follows in which the eyes are white, although a painless motility defect may be present. Clinical features broadly can be categorized into (i) soft tissue involvement, (ii) lid retraction, (iii) proptosis, (iv) optic neuropathy and (v) restrictive myopathy. A commonly used classification for the severity of TED has been issued by the European Group on Graves Orbitopathy (EUGOGO): (i) sight-threatening due to optic neuropathy or corneal breakdown; (ii) moderate–severe, with one of moderate–severe soft tissue involvement, lid retraction of 2 mm or more, diplopia and proptosis of 3 mm or more; (iii) mild, with only a minor impact on daily life.

Soft tissue involvement

- **Symptoms.** Grittiness, red eyes, lacrimation, photophobia, puffy lids and retrobulbar discomfort.
- **Signs** may include:
 - Epibulbar hyperaemia. This is a sensitive sign of inflammatory activity. Intense focal hyperaemia may outline the insertions of the horizontal recti (Fig. 3.7A).
 - Periorbital swelling is caused by oedema and infiltration behind the orbital septum; this may be associated with chemosis and prolapse of retroseptal fat into the eyelids (Fig. 3.7B).

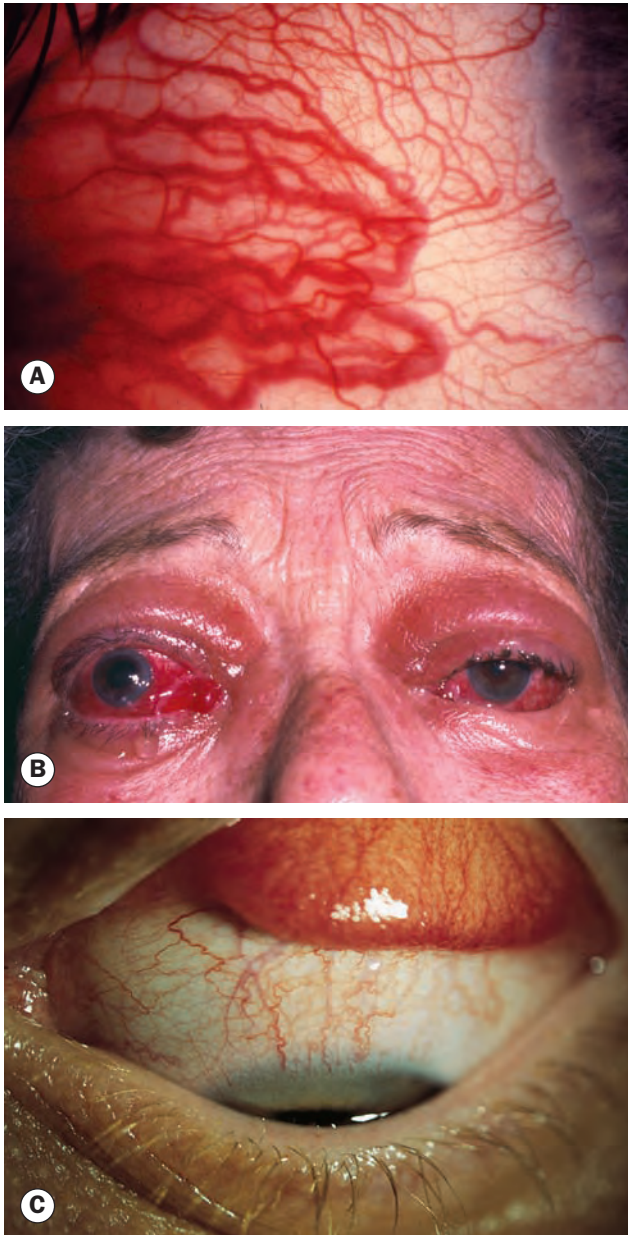


Fig. 3.7 Soft tissue involvement in thyroid eye disease. (A) Epibulbar hyperaemia overlying a horizontal rectus muscle; (B) periorbital oedema, chemosis and prolapse of fat into the eyelids; (C) superior limbic keratoconjunctivitis

- Tear insufficiency and instability is common.
- Corneal signs are exacerbated by lid retraction (see next) and can include punctate epithelial erosions, superior limbic keratoconjunctivitis (Fig. 3.7C and see Ch. 5), and occasionally bacterial keratitis, thinning and scarring.

Lid retraction

Retraction of upper and lower lids occurs in about 50% of patients with Graves disease. Humorally induced overaction of Müller muscle is postulated to occur as a result of sympathetic overstimulation secondary to high levels of thyroid hormones. Fibrotic

contracture of the levator palpebrae and inferior rectus muscles associated with adhesion to overlying orbital tissues is another probable mechanism, together with secondary overaction in response to hypo- or hypertropia produced by fibrosis.

- **Symptoms.** Patients may complain of a staring or bulging-eyed appearance, difficulty closing the eyes and ocular surface symptoms.
- **Signs**
 - The upper lid margin normally rests 2 mm below the limbus (Fig. 3.8A, right eye). Lid retraction is suspected when the margin is either level with or above the superior limbus, allowing sclera to be visible ('scleral show'; Fig. 3.8A, left eye).
 - The lower eyelid margin normally rests at the inferior limbus; retraction is suspected when sclera shows below the limbus. Lid retraction may occur in isolation or in association with proptosis, which exaggerates its severity.
 - The Dalrymple sign is lid retraction in primary gaze (Fig. 3.8B).
 - The Kocher sign describes a staring and frightened appearance of the eyes which is particularly marked on attentive fixation (Fig. 3.8C).
 - The von Graefe sign signifies retarded descent of the upper lid on downgaze (lid lag – Fig. 3.8D).

Proptosis

- **Symptoms** are similar to those of lid retraction.
- **Signs.** Proptosis is axial, unilateral or bilateral, symmetrical (Fig. 3.9A) or asymmetrical (Fig. 3.9B), and frequently permanent. Severe proptosis may compromise lid closure and along with lid retraction and tear dysfunction can lead to exposure keratopathy, corneal ulceration and infection (Fig. 3.9C).

Restrictive myopathy

Between 30% and 50% of patients with TED develop ophthalmoplegia and this may be permanent. Ocular motility is restricted initially by inflammatory oedema, and later by fibrosis.

- **Symptoms.** Double vision, and often discomfort in some positions of gaze.
- **Signs,** in approximate order of frequency:
 - Elevation defect (Fig. 3.10A) caused by fibrotic contracture of the inferior rectus, may mimic superior rectus palsy and is the most common motility deficit.
 - Abduction defect due to fibrosis of the medial rectus, which may simulate sixth nerve palsy.
 - Depression defect (Fig. 3.10B) secondary to fibrosis of the superior rectus.
 - Adduction defect caused by fibrosis of the lateral rectus.

Optic neuropathy

Optic neuropathy is a fairly common (up to 6%) serious complication caused by compression of the optic nerve or its blood supply at the orbital apex by the congested and enlarged recti (Fig. 3.11) and swollen orbital tissue. Such compression, which may

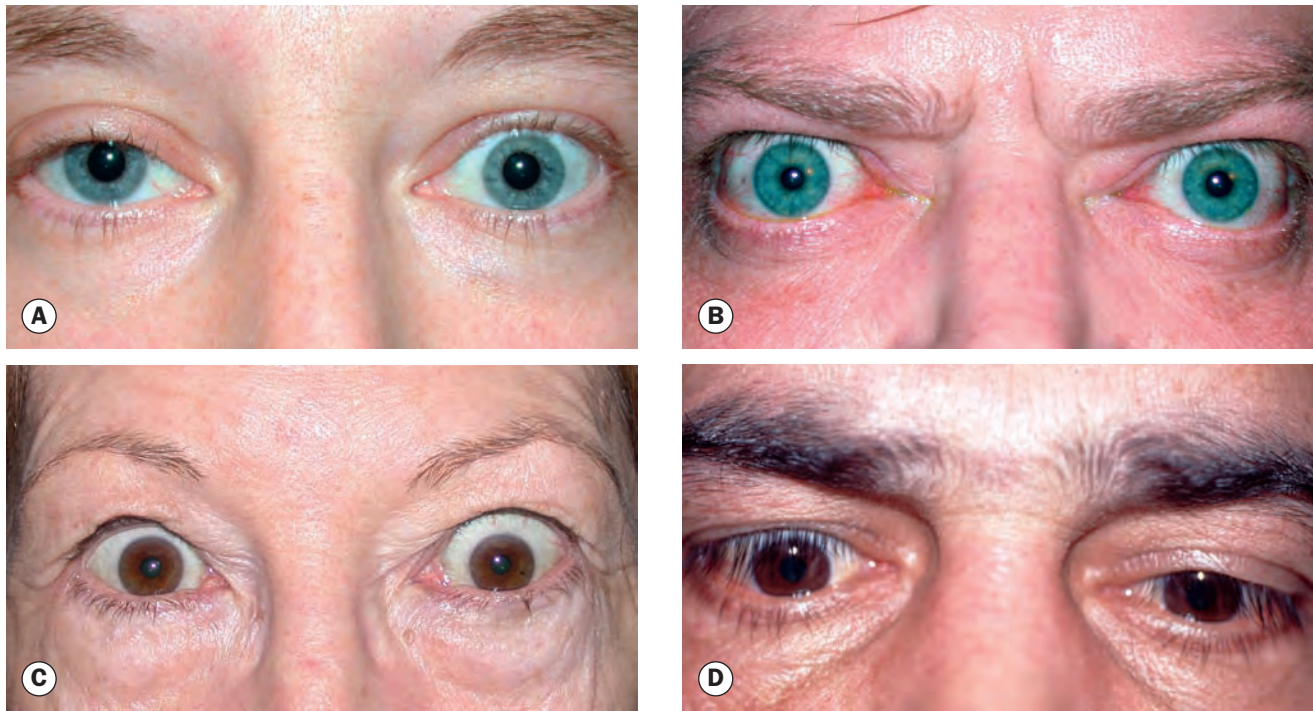


Fig. 3.8 Lid signs in thyroid eye disease. (A) Mild left lid retraction; (B) moderate bilateral symmetrical lid retraction – Dalrymple sign; (C) severe bilateral lid retraction – Kocher sign; (D) right lid lag on downgaze – von Graefe sign

(Courtesy of G Rose – fig. B; S Chen – fig. C)

occur in the absence of significant proptosis, may lead to severe visual impairment if adequate and timely treatment is not instituted.

- **Symptoms.** Impairment of central vision occurs in conjunction with other symptoms of TED. In order to detect early involvement, patients should be advised to monitor their own visual function by alternately occluding each eye, reading small print and assessing the intensity of colours, for example on a television screen.
- **Signs.** A high index of suspicion should be maintained for optic neuropathy, and it is important not to mistakenly attribute disproportionate visual loss to minor disease.
 - Visual acuity (VA) is usually reduced, but not invariably.
 - Colour desaturation is a sensitive feature.
 - There may be diminished light brightness appreciation.
 - A relative afferent pupillary defect, if present, should give cause for marked concern.
 - Visual field defects can be central or paracentral and may be combined with nerve fibre bundle defects. These findings, in concert with elevated IOP, may be confused with primary open-angle glaucoma.
 - The optic disc may be normal, swollen or, rarely, atrophic.

Investigation

Investigations other than blood tests for thyroid disease are not necessary if the diagnosis is evident clinically, but the exclusion of other conditions is sometimes indicated. Visual field testing is carried out if there is a suspicion of optic nerve compromise, and

may be performed as part of a baseline evaluation even if there is no apparent visual impairment. MRI, CT and ultrasonographic imaging of the orbits are indicated in some circumstances, such as helping to confirm an equivocal diagnosis by identification of the typical pattern of extraocular muscle involvement in TED, consisting of muscle belly enlargement with tendon sparing. Imaging is also used in the assessment of optic nerve compression and prior to orbital wall surgery. Visual evoked potentials are sometimes utilized in optic neuropathy.

Treatment

Treatment can be classified into that of mild disease (most patients), moderate to severe active disease, and treatment of post-inflammatory complications. The first measure taken in all cases should be the cessation of smoking. Thyroid dysfunction should also be managed adequately; if radioiodine treatment is administered in patients with pre-existing TED, a short course of oral steroids should be given in concert.

- **Mild disease**
 - Lubricants for superior limbic keratoconjunctivitis, corneal exposure and dryness.
 - Topical anti-inflammatory agents (steroids, non-steroidal anti-inflammatory drugs (NSAIDs), ciclosporin) are advocated by some authorities.
 - Head elevation with three pillows during sleep to reduce periorbital oedema.
 - Eyelid taping during sleep may alleviate mild exposure keratopathy.

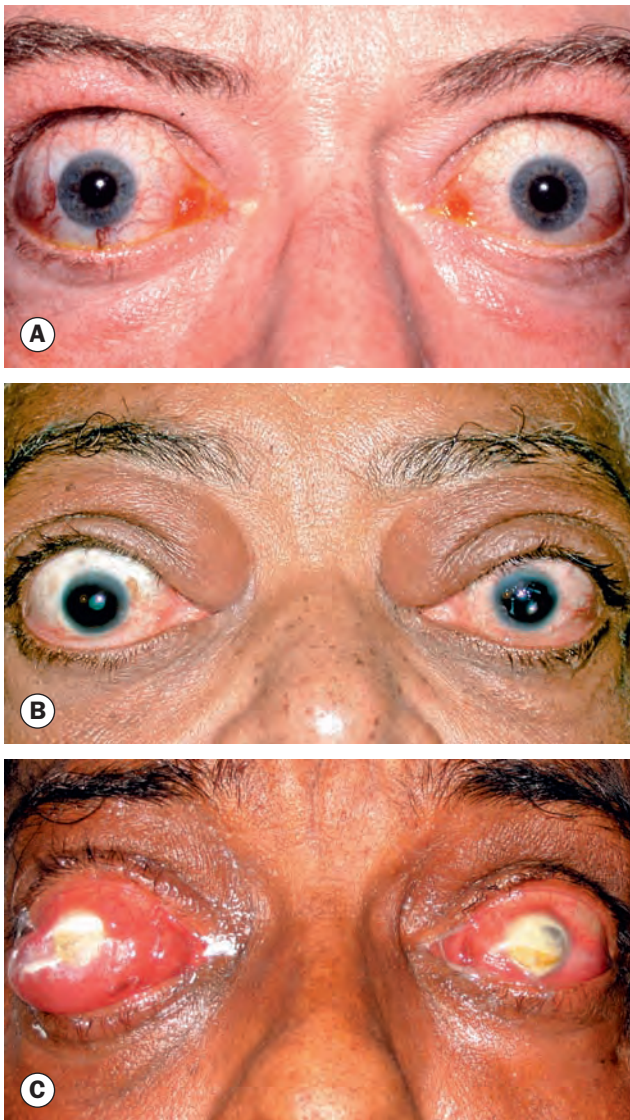


Fig. 3.9 Proptosis in thyroid eye disease. (A) Symmetrical; (B) asymmetrical; (C) bacterial keratitis due to severe exposure

(Courtesy of A Pearson – figs A and B; S Kumar Puri – fig. C)

- **Moderate to severe active disease**

- Clinical activity score. EUGOGO suggests calculating a ‘clinical activity score’ to aid in determining a threshold for the use of immunosuppressives, assigning one point for each feature present from the following list and considering treatment for a score of 3 or more out of 7.
 1. Spontaneous orbital pain.
 2. Gaze-evoked orbital pain.
 3. Eyelid swelling considered to be due to active (inflammatory phase) TED.
 4. Eyelid erythema.
 5. Conjunctival redness considered to be due to active (inflammatory phase) TED.

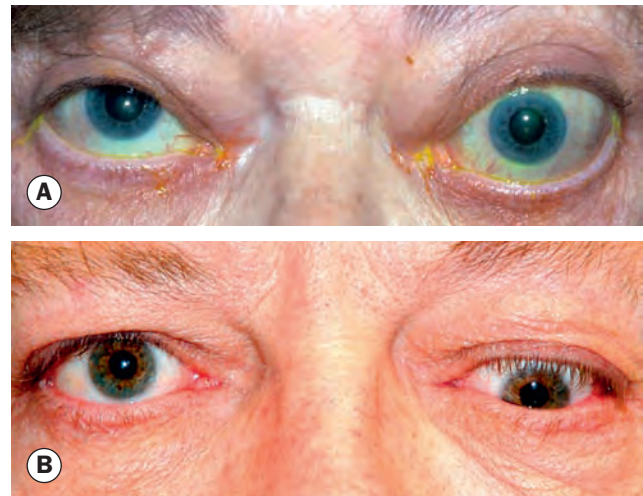


Fig. 3.10 Restrictive thyroid myopathy. (A) Defective elevation of the left eye; (B) defective depression of the right eye

6. Chemosis.

7. Inflammation of caruncle or plica.

During subsequent review, a point is allocated for an increase in proptosis of 2 mm or more, a decrease in uniocular excursion in any one direction of 8° or more, or a decrease in Snellen acuity of one line.

- Systemic steroids are the mainstay of treatment for moderate to severe disease. Oral prednisolone 60–80 mg/day may be given initially, and tapered depending on response. Intravenous methylprednisolone is often reserved for acute compressive optic neuropathy (see below), but tolerability is better and outcomes may be superior compared with oral treatment; a lower-intensity regimen in the absence of acute sight-threatening disease is 0.5 g once weekly for 6 weeks followed by 0.25 g once weekly for 6 weeks. A reduction in discomfort, chemosis and periorbital oedema usually occurs within 24 hours, with a maximal response within 2–8 weeks. Ideally, oral steroid therapy should be discontinued after several months, but long-term low-dose maintenance may be necessary.
- Orbital steroid injections are occasionally used in selected cases to minimize systemic side effects, but are typically considerably less effective than systemic treatment.
- Low-dose fractionated radiotherapy may be used in addition to steroids or when steroids are contraindicated or ineffective, but because of the delayed effect is not used as the sole treatment of acute optic nerve compression. A positive response is usually evident within 6 weeks, with maximal improvement by 4 months; around 40% will not respond. Adverse effects include cataract, radiation retinopathy, optic neuropathy and an increased risk of local cancer; the threshold for its use should be higher in younger patients and diabetics, the latter because of a possibly increased risk of retinopathy.

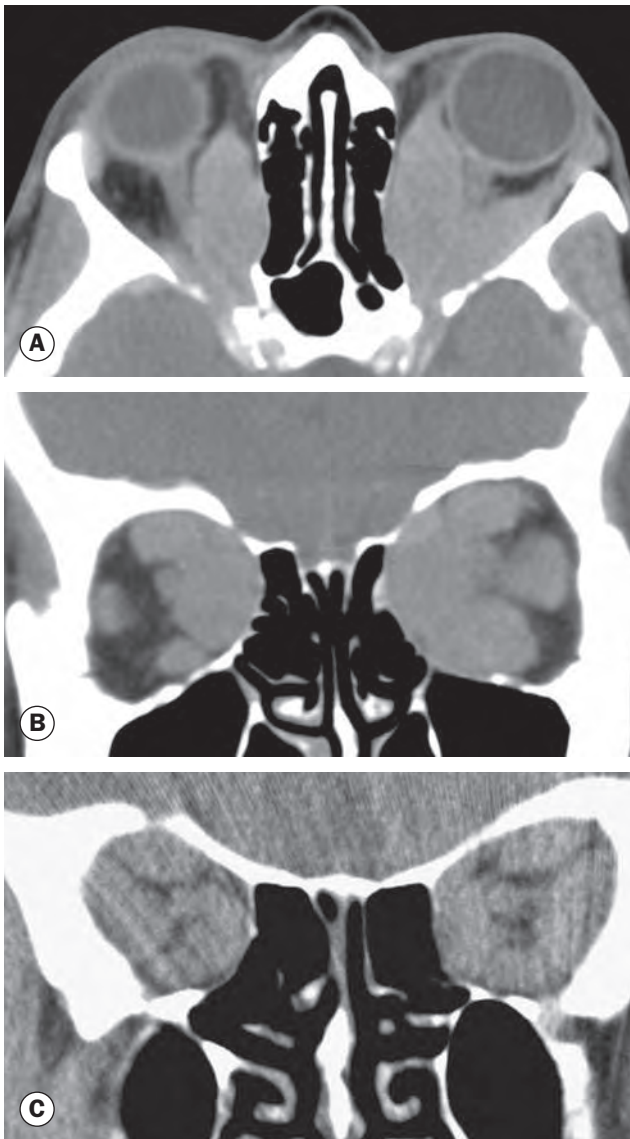


Fig. 3.11 CT shows muscle enlargement in thyroid eye disease. (A) Axial view; (B) coronal view – note sparing of the right lateral rectus muscle; (C) coronal view shows crowding at the orbital apex

(Courtesy of N Sibtain – figs A and B; J Nerad, K Carter and M Alford, from 'Oculoplastic and Reconstructive Surgery', in *Rapid Diagnosis in Ophthalmology*, Mosby 2008 – fig. C)

- Combined therapy with irradiation, azathioprine and low-dose prednisolone may be more effective than steroids or radiotherapy alone.
- Optic neuropathy, and less commonly intractable corneal exposure, requires aggressive treatment. Pulsed intravenous methylprednisolone is commonly used, regimens including 0.5–1 g on three successive days with conversion to oral treatment (e.g. 40 mg/day prednisolone) or 0.5–1 g on alternate days, 3–6 times, keeping the maximum dose below 8 g to reduce the risk of liver compromise, followed by oral prednisolone; appropriate monitoring should be instituted, including

liver function tests, as well as gastric protective treatment and osteoporosis prophylaxis if necessary. Orbital wall decompression (see below) and/or orbital apex decompression may be considered if steroids are ineffective (20% receiving intravenous treatment) or contraindicated. Orbital radiotherapy may also be administered, but is generally only used as an adjunct to other modalities.

- Several drugs targeting specific aspects of the immune response in TED are under investigation, notably monoclonal antibody treatment with rituximab.
- **Post-inflammatory complications.** Eyelid surgery should be performed only after any necessary orbital and then strabismus procedures have been undertaken, as orbital decompression may impact both ocular motility and eyelid position, and extraocular muscle surgery may affect eyelid position.
 - Proptosis. After active inflammation has remitted, the patient can be left with cosmetically and functionally significant proptosis, the treatment of which is essentially surgical. Surgical decompression increases the volume of the orbit by removing the bony walls and may be combined with removal of orbital fat. Most surgery is undertaken via an external approach, though the medial wall and the medial part of the floor can be reached endoscopically. One-wall (deep lateral) decompression is effective (approximately 4–5 mm reduction in proptosis) and may reduce the risk of postoperative diplopia; two-wall (balanced medial and lateral – Fig. 3.12) decompression provides a greater effect but with a significant risk of inducing diplopia; three-wall decompression includes the floor with a reduction in proptosis of 6–10 mm but may lead to hypoglobus and carries a higher risk of infraorbital nerve damage and diplopia; very severe proptosis may require removal of part of the orbital roof in addition (four-wall decompression).
 - Restrictive myopathy. Surgery is required in most cases experiencing persistent diplopia in the primary or reading



Fig. 3.12 Axial CT following bilateral lateral and medial wall decompression

(Courtesy of A Pearson)

positions of gaze, provided the inflammatory stage has subsided and the angle of deviation has been stable for at least 6–12 months. Until these criteria are met, diplopia may be alleviated, if possible, with prisms or sometimes botulinum toxin. The goal of operative treatment is to achieve binocular single vision in the primary and reading positions; restrictive myopathy often precludes binocularity in all positions of gaze, though with time the field of binocular single vision may enlarge as a result of increasing fusional vergence. Recession of the inferior and/or medial recti is the most commonly indicated surgery (a rectus muscle is never resected, only recessed in TED), generally utilizing adjustable sutures (see Ch. 18). The suture is adjusted later the same day or on the first postoperative day to achieve optimal alignment, and the patient is encouraged subsequently to practise achieving single vision with a consistently accessible target such as a television.

- Lid retraction. Mild lid retraction frequently improves spontaneously so does not require treatment. Control of hyperthyroidism may also be beneficial. Botulinum toxin injection to the levator aponeurosis and Müller muscle may be used as a temporary measure in patients awaiting definitive correction. Müllerotomy (disinsertion of Müller muscle) is effective for mild lid retraction, but more severe cases may also require recession/disinsertion of the levator aponeurosis and the suspensory ligament of the superior conjunctival fornix. Recession of the lower lid retractors, with or without a hard palate graft, can be used when retraction of the lower lid is 2 mm or more (see also Ch. 1).

INFECTIONS

Preseptal cellulitis

Introduction

Preseptal cellulitis is an infection of the subcutaneous tissues anterior to the orbital septum. It is considerably more common than orbital cellulitis, and though regarded as less serious, can still be associated with severe complications such as abscess formation, meningitis and cavernous sinus thrombosis. Rapid progression to orbital cellulitis may occasionally occur. Organisms typically responsible are *Staphylococcus aureus* and *Streptococcus pyogenes*, with causes including skin trauma such as laceration or insect bites, spread from focal ocular or periocular infection such as an acute hordeolum, dacryocystitis, conjunctivitis or sinusitis, and haematogenous spread from remote infection such as the upper respiratory tract or middle ear.

Diagnosis

The condition manifests with a swollen, often firm, tender red eyelid that may be very severe (Fig. 3.13A); however, in contrast to



Fig. 3.13 Preseptal cellulitis. (A) Left preseptal cellulitis resulting from an infected eyelid abrasion; (B) axial CT shows opacification anterior to the orbital septum (Courtesy of C Barry – fig. A)

orbital cellulitis, proptosis and chemosis are absent, and visual acuity, pupillary reactions and ocular motility are unimpaired. The patient is often pyrexial. Imaging with MRI or CT (Fig. 3.13B) is not indicated unless orbital cellulitis or a lid abscess is suspected, or there is a failure to respond to therapy.

Treatment

Treatment is with oral antibiotics such as co-amoxiclav 250–500 mg/125 mg 2–3 times daily or 875/125 mg twice daily, depending on severity. Severe infection may require intravenous antibiotics. The patient's tetanus status should be ascertained in cases following trauma.

Bacterial orbital cellulitis

Introduction

Bacterial orbital cellulitis is a serious infection of the soft tissues behind the orbital septum, which can be sight- and life-threatening. It can occur at any age but is more common in children. *Streptococcus pneumoniae*, *Staphylococcus aureus*, *Streptococcus pyogenes* and *Haemophilus influenzae* are common causative organisms, with infection originating typically from the paranasal (especially ethmoid) sinuses. Infection can also spread from preseptal cellulitis, dacryocystitis, midfacial skin or dental infection, and can

follow trauma, including any form of ocular surgery. Blood-borne spread from infection elsewhere in the body may occur.

Clinical features

- **Symptoms** consist of the rapid onset of pain exacerbated by eye movement, swelling of the eye, malaise, and frequently visual impairment and double vision. There is commonly a recent history of nasal, sinus or respiratory symptoms.
- **Signs**
 - Pyrexia, often marked.
 - VA may be reduced and colour vision impaired, raising the possibility of optic nerve compression; the presence of a relative afferent pupillary defect in a previously normal eye makes this almost certain.
 - Tender, firm, erythematous and warm eyelids, with periocular and conjunctival (chemosis) oedema, conjunctival injection and sometimes subconjunctival haemorrhage; the signs are usually unilateral, though oedema may spread to the contralateral eyelids.
 - Proptosis is common in established infection, but is often obscured by lid swelling; it may be non-axial (dystopia), particularly if an abscess is present.
 - Painful ophthalmoplegia (Fig. 3.14A).

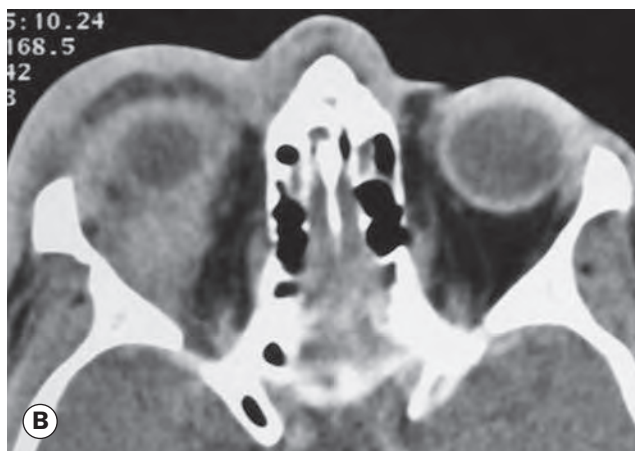


Fig. 3.14 (A) Right orbital cellulitis with ophthalmoplegia; (B) axial CT shows both preseptal and orbital opacification

Table 3.1 Differential diagnosis of an acutely inflamed orbit

Infection
• Bacterial orbital cellulitis • Fungal orbital infection • Dacryocystitis • Infective dacryoadenitis
Vascular lesions
• Acute orbital haemorrhage • Cavernous sinus thrombosis • Carotid-cavernous fistula
Neoplasia
• Rapidly progressive retinoblastoma • Lacrimal gland tumour • Other neoplasm, e.g. metastatic lesion with inflammation, lymphoma, Waldenström macroglobulinaemia • Rhabdomyosarcoma, leukaemia, lymphangioma or neuroblastoma in children
Endocrine
• Thyroid eye disease of rapid onset
Non-neoplastic inflammation
• Idiopathic orbital inflammatory disease • Tolosa-Hunt syndrome • Orbital myositis • Acute allergic conjunctivitis with lid swelling • Herpes zoster ophthalmicus • Herpes simplex skin rash • Sarcoidosis • Vasculitides: Wegener granulomatosis, polyarteritis nodosa • Scleritis, including posterior scleritis • Ruptured dermoid cyst

- Choroidal folds and optic disc swelling may be present on fundus examination.
- **Differential diagnosis.** Major diagnostic alternatives are listed in Table 3.1.
- **Complications**
 - Ocular complications include optic neuropathy, exposure keratopathy, raised IOP, endophthalmitis and occlusion of the central retinal artery or vein.
 - Subperiosteal abscess, most frequently located along the medial orbital wall.
 - Intracranial complications, which are uncommon (3–4%) but extremely serious, include meningitis, brain abscess and cavernous sinus thrombosis.

Investigation

Investigations may include:

- Ascertainment of tetanus immunization status in cases of trauma.
- White cell count.
- Blood cultures.
- Culture of nasal discharge.
- High-resolution CT of the orbit, sinuses and brain (Fig. 3.14B) is vital to confirm the diagnosis and exclude a

subperiosteal or intracranial abscess. MRI is also sometimes performed.

- Lumbar puncture if meningeal or cerebral signs develop.

Treatment

- **Hospital admission** is mandatory, with urgent otolaryngological assessment and frequent ophthalmic review. Paediatric specialist advice should be sought in the management of a child, and a low threshold should be adopted for infectious disease specialist consultation.
- **Delineation** of the extent of erythema on the skin using a surgical marker may help in judging progress.
- **Antibiotics** are given intravenously, with the specific drug depending on local sensitivities; ceftazidime is a typical choice, supplemented by oral metronidazole to cover anaerobes. Intravenous antibiotics should be continued until the patient has been afebrile for 4 days, followed by 1–3 weeks of oral treatment.
- **Monitoring of optic nerve function** is performed at least every 4 hours initially by testing VA, colour vision, light brightness appreciation and pupillary reactions. Deterioration should prompt the consideration of surgical intervention.
- **Surgery.** Drainage of an orbital abscess should be considered at an early stage; drainage of infected sinuses should be considered if there is a lack of response to antibiotics, or if there is very severe sinus disease. Biopsy of inflammatory tissue may be performed for an atypical clinical picture. Severe optic nerve compression may warrant an emergency canthotomy/cantholysis (see Ch. 21).

Rhino-orbital mucormycosis

Introduction

Mucormycosis is a rare aggressive and often fatal infection caused by fungi of the family Mucoraceae. It typically affects patients with diabetic ketoacidosis or immunosuppression and is extremely rare in the immunocompetent. Infection is acquired by the inhalation of spores, which give rise to an upper respiratory infection. Spread then occurs to the contiguous sinuses and subsequently to the orbit and brain. Invasion of blood vessels by the hyphae results in occlusive vasculitis with infarction of orbital tissues.

Diagnosis

- **Symptoms.** Gradual onset facial and periorbital swelling, diplopia and visual loss.
- **Signs** are similar to bacterial orbital cellulitis, but tend to be less acute and with slower progression. Infarction superimposed on septic necrosis is responsible for the classic black eschar that may develop on the palate, turbinates, nasal septum, skin and eyelids (Fig. 3.15).
- **Complications** include retinal vascular occlusion, multiple cranial nerve palsies and cerebrovascular occlusion.
- **Differential diagnosis** is listed in Table 3.1.



Fig. 3.15 Necrosis of the eyelid in rhino-orbital mucormycosis

- **Investigation** is much the same as for bacterial orbital cellulitis.

Treatment

- Correction of the underlying metabolic defect should be instituted if possible.
- Intravenous antifungal treatment.
- Daily packing and irrigation of the involved areas with antifungal agent.
- Wide excision of devitalized and necrotic tissues; exenteration may be required in unresponsive cases in order to reduce the risk of death.
- Adjunctive hyperbaric oxygen may be helpful.

NON-INFECTIVE INFLAMMATORY DISEASE

Idiopathic orbital inflammatory disease

Idiopathic orbital inflammatory disease (IOID; also non-specific orbital inflammation or orbital pseudotumour) is an uncommon disorder characterized by non-neoplastic, non-infective, space-occupying orbital infiltration with inflammatory features. The process may preferentially involve any or all of the orbital soft tissues. Histopathological analysis reveals pleomorphic inflammatory cellular infiltration followed by reactive fibrosis. Unilateral disease is typical in adults, although in children bilateral involvement may occur. Intracranial extension is rare; simultaneous orbital and sinus involvement is also rare, and may be a distinct entity.

Diagnosis

- **Symptoms** typically consist of acute or subacute ocular and periocular redness, swelling and pain (Fig. 3.16A). Systemic symptoms are common in children.

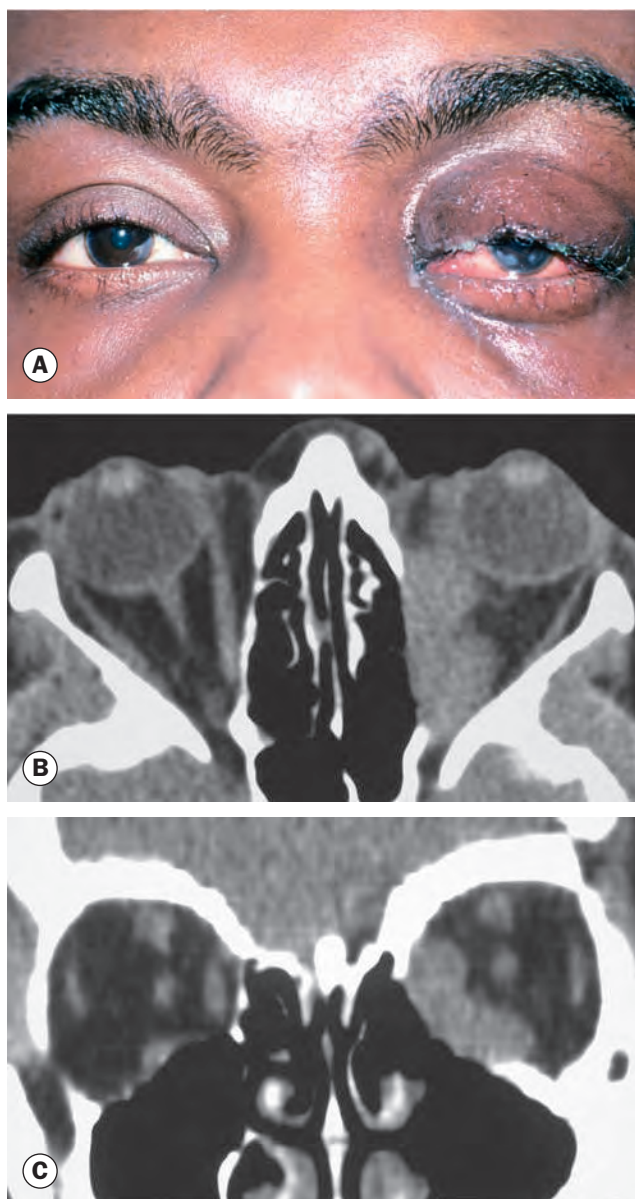


Fig. 3.16 (A) Left idiopathic orbital inflammatory disease; (B) CT axial view shows ill-defined orbital opacification; (C) coronal view

(Courtesy of R Bates – fig. A; A Pearson – figs B and C)

• Signs

- Pyrexia is present in up to 50% of children, but is rare in adults.
- Congestive proptosis.
- Mild to severe ophthalmoplegia may occur.
- Features of optic nerve dysfunction, particularly if the inflammation involves the posterior orbit; there may be optic disc swelling.
- Choroidal folds, if present, may be associated with reduced vision but optic neuropathy must always be suspected.

- **Course.** The natural history of the inflammatory process is very variable.
 - Spontaneous remission after a few weeks without sequelae.
 - Intermittent episodes of activity, usually with eventual remission.
 - Severe prolonged inflammation eventually leading to progressive fibrosis of orbital tissues, resulting in a 'frozen orbit' characterized by ophthalmoplegia, which may be associated with ptosis and visual impairment caused by optic nerve involvement.
- **Investigation**
 - CT shows ill-defined orbital opacification and loss of definition of contents (Figs 3.16B and C).
 - Biopsy is generally required in persistent cases to confirm the diagnosis, and particularly to rule out neoplasia and systemic inflammatory conditions.
 - A wide range of other investigations may be considered to aid in the exclusion of alternative diagnoses, particularly infection, lymphoma and non-neoplastic infiltrative disorders such as sarcoidosis and Wegener granulomatosis.

Treatment

- **Observation**, for relatively mild disease, in anticipation of spontaneous remission.
- **NSAIDs** alone (e.g. ibuprofen) are often effective and may be tried in mild disease prior to steroid therapy. Co-prescription of a proton pump inhibitor should be considered.
- **Systemic steroids** should be administered only after the diagnosis has been confirmed, as they may mask other pathology such as infection and Wegener granulomatosis. Oral prednisolone is initially given at a dose of 1.0–1.5 mg/kg/day, subsequently being tapered and discontinued over a number of weeks depending on clinical response; further treatment may be needed in the event of recurrence.
- **Orbital depot steroid** injection may be useful in some cases.
- **Radiotherapy** may be considered if there has been no improvement after 2 weeks of adequate steroid therapy. Even low-dose treatment (e.g. 10 Gy) may produce remission, though much higher total doses may be necessary.
- **Other options**, usually as supplementary treatments or in resistant cases, include cytotoxic drugs (e.g. methotrexate, azathioprine), calcineurin inhibitors (e.g. ciclosporin, tacrolimus) and biological blockers.
- **Surgical resection** of an inflammatory focus may be contemplated in highly resistant cases.

Orbital myositis

Introduction

Orbital myositis is an idiopathic, non-specific inflammation of one or more extraocular muscles and is considered a subtype of

IOID. Histology shows a chronic inflammatory cellular infiltrate associated with the muscle fibres (Fig. 3.17A).

Diagnosis

- **Symptoms.** Acute pain, exacerbated by eye movement, and diplopia; onset is usually in early adulthood.
- **Signs** are generally more subtle than IOID.
 - Lid oedema, ptosis and chemosis.
 - Pain and diplopia associated with eye movements.
 - Vascular injection over the involved muscle (Fig. 3.17B).
 - In chronic cases the affected muscle may become fibrosed, with permanent restrictive myopathy.
- **Course**
 - Acute non-recurrent involvement that resolves spontaneously within 6 weeks.
 - Chronic disease characterized by either a single episode persisting for longer than 2 months (often for years) or recurrent attacks.
- **Investigation** consists primarily of MRI or CT, which show enlargement of the affected muscles (Fig. 3.17C), with or without involvement of the tendons of insertion; this is in contrast to TED-related muscle enlargement, in which the tendon is always spared. Additional investigations may be required in some cases.

Treatment

Treatment is aimed at relieving discomfort and dysfunction, shortening the course and preventing recurrences. NSAIDs may be adequate in mild disease, but systemic steroids are generally required and usually produce dramatic improvement, although recurrence is seen in 50%. Radiotherapy is also effective, particularly in limiting recurrence.

Acute dacryoadenitis

Acute dacryoadenitis may be idiopathic or due to viral (e.g. mumps, Epstein–Barr, cytomegalovirus) or – rarely – bacterial infection; the lacrimal gland is often involved in IOID. Chronic conditions such as sarcoidosis, Sjögren syndrome, thyroid disease and some chronic infections usually give a less acute onset, and involvement can be bilateral. Presentation in acute disease is with the rapid onset of discomfort in the region of the gland. Lacrimal secretion may be reduced or increased, and discharge may be reported. Swelling of the lateral aspect of the eyelid overlying the palpebral lobe leads to a characteristic S-shaped ptosis, and enlargement of the orbital lobe may give a slight downward and inward dystopia (Fig. 3.18A) and occasionally proptosis and other signs of orbital disease. There is tenderness over the lacrimal gland, and injection of the conjunctiva overlying the palpebral lobe may be seen on upper lid eversion (Fig. 3.18B). Chemosis may be present. There may be local (e.g. pre-auricular) lymph node enlargement. CT shows enlargement of the gland and involvement of adjacent tissues (Fig. 3.18C) without bony erosion; the latter suggests a tumour. Biopsy is sometimes indicated, particularly to

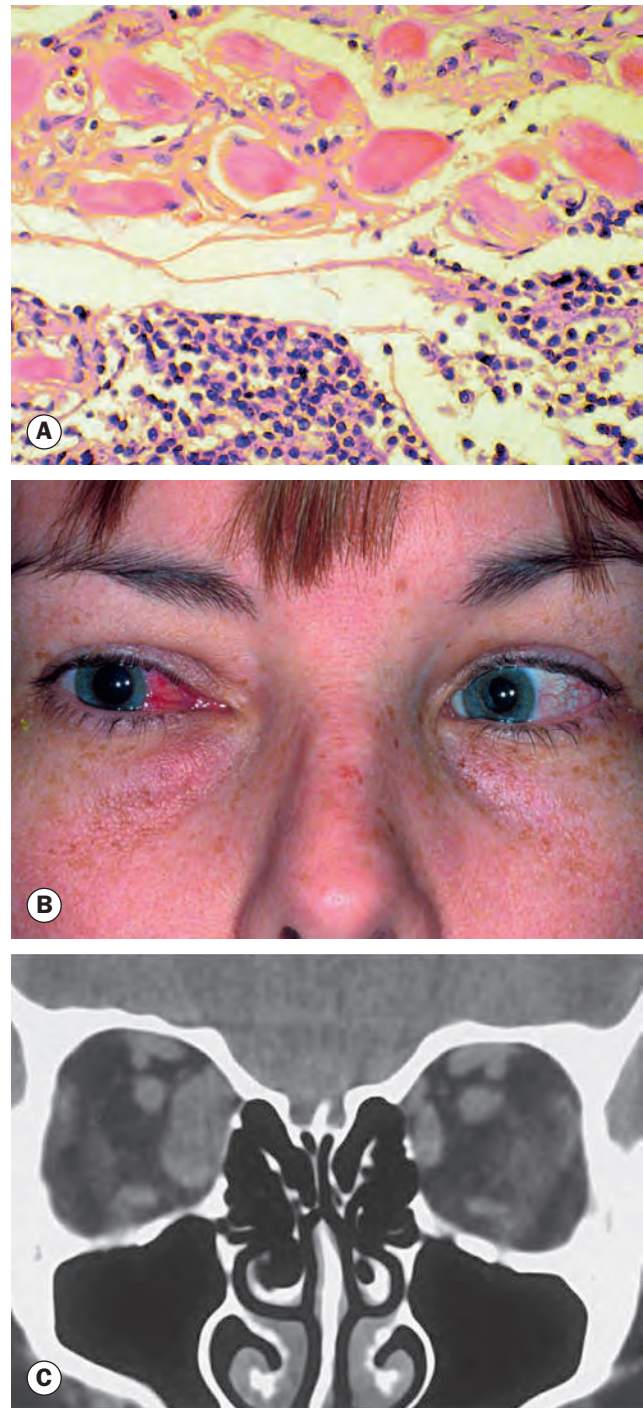


Fig. 3.17 Orbital myositis. (A) Histology shows a chronic inflammatory cellular infiltrate in relation to muscle fibres; (B) vascular injection over the insertion of the right medial rectus; (C) coronal CT shows enlargement of the right medial rectus

(Courtesy of J Harry and G Misson, from *Clinical Ophthalmic Pathology*, Butterworth-Heinemann 2001 – fig. A; J Nerad, K Carter and M Alford, from *Oculoplastic and Reconstructive Surgery*, in *Rapid Diagnosis in Ophthalmology*, Mosby 2008 – figs B and C)

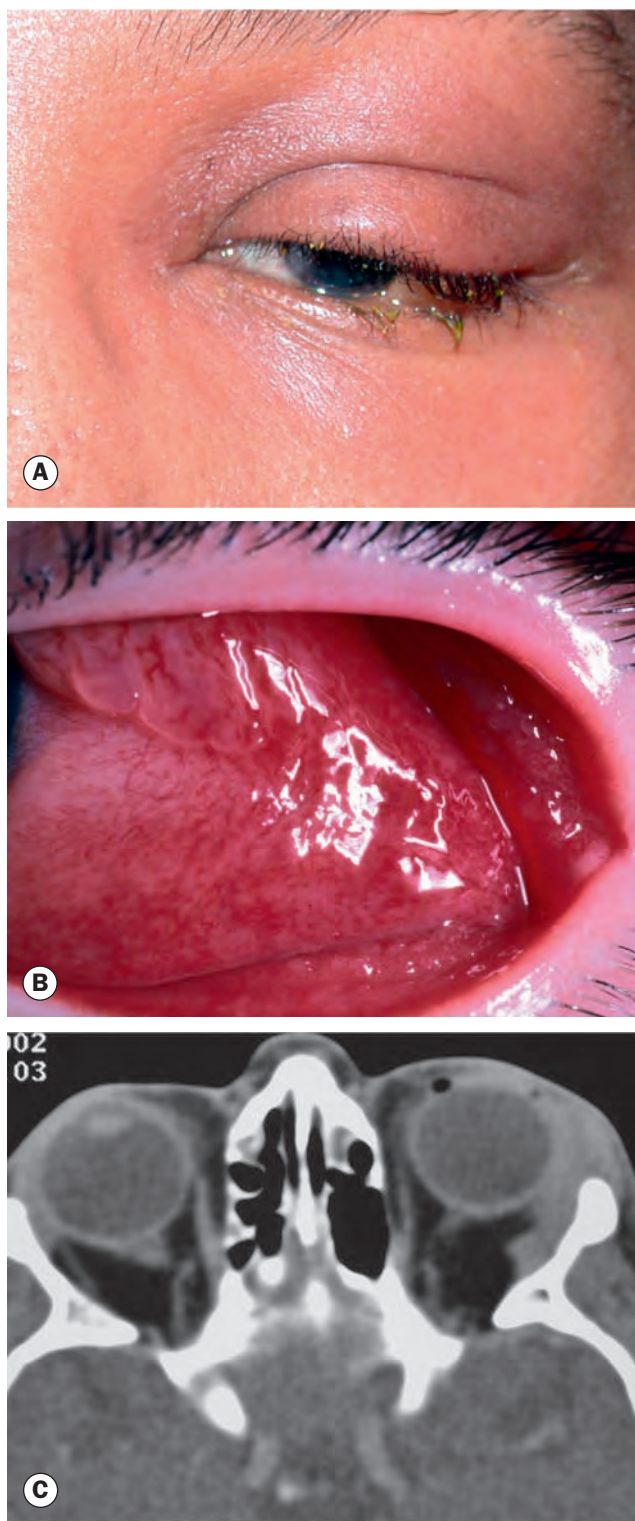


Fig. 3.18 Left acute dacryoadenitis. **(A)** Swelling on the lateral aspect of the eyelid and an S-shaped ptosis; **(B)** injection of the palpebral portion of the lacrimal gland and adjacent conjunctiva; **(C)** axial CT shows enlargement of the gland and opacification of adjacent tissues

(Courtesy of R Bates – fig. B; A Pearson – fig. C)

exclude a tumour. Treatment varies according to the cause, but in many cases is not required.

Tolosa–Hunt syndrome

Tolosa–Hunt syndrome is a rare idiopathic condition caused by non-specific granulomatous inflammation of the cavernous sinus, superior orbital fissure and/or orbital apex. It is a diagnosis of exclusion and should be investigated fully. Presentation is with ipsilateral periorbital or hemicranial pain, and diplopia due to one or more ocular motor pareses, with pupillary and eyelid involvement in many cases. Proptosis, if present, is usually mild. Sensory loss along the distribution of the first and second divisions of the trigeminal nerve is common. The patient may be pyrexial. Diagnosis is with imaging, together with other investigations to rule out identifiable causes, including neoplasia. Treatment is with systemic steroids and other immunosuppressants as necessary; the clinical course is characterized by remissions and recurrences.

Wegener granulomatosis

Wegener granulomatosis (see Ch. 8), an idiopathic multisystem granulomatous disorder that may involve the orbit, often bilaterally, usually by contiguous spread from the paranasal sinuses or nasopharynx. Primary orbital involvement is less common. The possibility of Wegener granulomatosis should be considered in any patient with bilateral orbital inflammation, particularly if associated with sinus pathology. Antineutrophilic cytoplasmic antibody (cANCA variant) is a useful serological test. Other ocular features include scleritis, peripheral ulcerative keratitis, intraocular inflammation and retinal vascular occlusions. Treatment is with cyclophosphamide and steroids, which are usually effective. In resistant cases ciclosporin, azathioprine, antithymocyte globulin or plasmapheresis may be useful. Surgical decompression may be required for severe orbital involvement.

NON-NEOPLASTIC VASCULAR ABNORMALITIES

Cavernous sinus thrombosis

This refers to clotting within the cavernous sinus, usually resulting from infection such as sinusitis, orbital or preseptal cellulitis or otitis. There is a high mortality rate: 20% treated and up to 100% untreated. Features are of rapid onset and may include severe headache, malaise, nausea and vomiting, unilateral or often bilateral proptosis, chemosis, congestion of the facial, conjunctival and retinal veins, reduced vision, and signs resulting from compromised function of the third to sixth cranial nerves, which run through the cavernous sinus. Diagnosis is with imaging, especially MRI and MRI venography; systemic investigation for infection is also performed, including lumbar puncture. Treatment consists of intravenous antibiotics and sometimes surgical drainage.

Carotid–cavernous fistula

Introduction

A carotid–cavernous fistula involves the development of an arteriovenous fistula between the carotid artery and the venous cavernous sinus (see Fig. 19.60) with a rise in venous pressure in the sinus and structures draining to it. Ocular manifestations occur because of venous and arterial stasis around the eye and orbit, increased episcleral venous pressure and a decrease in arterial blood flow to the cranial nerves within the cavernous sinus. Carotid–cavernous fistulae are classified into ‘direct’ and ‘indirect’ forms.

- Direct fistulae are high-flow shunts in which carotid artery blood passes directly into the cavernous sinus through a defect in the wall of the intracavernous portion of the internal carotid artery as a result of trauma (75%), including surgery, spontaneous rupture of an intracavernous carotid aneurysm or an atherosclerotic artery, the latter frequently in a middle-aged hypertensive woman; spontaneous fistulae usually have lower flow.
- In an indirect fistula (‘dural shunt’), the intracavernous portion of the internal carotid artery remains intact. Arterial blood flows through the meningeal branches of the external or internal carotid arteries indirectly into the cavernous sinus, and the clinical features are more subtle than in a direct fistula such that the condition may be overlooked. Spontaneous rupture of an atherosclerotic artery or of a congenital malformation is the usual cause, and may be precipitated by minor trauma or straining. Connective tissue and collagen vascular disorders can be associated.

Diagnosis

- **Symptoms – direct.** Presentation may be days or weeks after head injury with a classic triad of pulsatile proptosis, conjunctival chemosis and a whooshing noise in the head.
- **Symptoms – indirect.** Gradual onset of redness of one or both eyes is a typical presentation, caused by conjunctival vascular engorgement.
- **Signs – direct**
 - Immediate visual loss may be due to ocular or optic nerve damage at the time of head trauma.
 - Delayed visual loss may occur as a result of exposure keratopathy, secondary glaucoma, central retinal vein occlusion, anterior segment ischaemia or ischaemic optic neuropathy.
 - Signs are usually ipsilateral to the fistula but may be bilateral, or even contralateral, because of midline connections between the two cavernous sinuses.
 - Marked epibulbar vascular dilatation (Fig. 3.19A).
 - Chemosis, commonly haemorrhagic, particularly in the early stages (Fig. 3.19B).
 - Pulsatile proptosis associated with a bruit and a thrill, both of which can be abolished by ipsilateral carotid compression in the neck.
- Increased IOP due to elevated episcleral venous pressure and orbital congestion, and sometimes angle-closure glaucoma.
- Anterior segment ischaemia, characterized by corneal epithelial oedema, aqueous cells and flare, and in severe cases iris atrophy, cataract and rubeosis iridis.
- Ptosis due to third nerve involvement.
- Ophthalmoplegia (60–70%) due to the ocular motor nerve damage from initial trauma, an intracavernous aneurysm or the fistula itself. The sixth cranial nerve is most frequently affected because of its free-floating location within the cavernous sinus. The third and fourth nerves, situated in the lateral wall of the sinus, are less frequently involved. Engorgement and swelling of extraocular muscles may also contribute to defective ocular motility.
- Fundus examination may show optic disc swelling, venous dilatation and intraretinal haemorrhages (Fig. 3.19C) from venous stasis and impaired retinal blood flow. Vitreous haemorrhage is rare.
- **Signs – indirect**
 - Milder epibulbar vascular dilatation than with a direct fistula (Fig. 3.19D).
 - Exaggerated ocular pulsation; this is readily detected on slit lamp applanation tonometry.
 - The presence of ‘corkscrew’ epibulbar vessels (Fig. 3.19E) is a common subtle later sign; these are not pathognomonic and can be found in normal eyes.
 - Raised IOP, often bilateral but higher on the side of the fistula.
 - Proptosis and bruit are mild if present.
 - Ophthalmoplegia caused by sixth nerve palsy or swelling of extraocular muscles in marked cases.
 - Fundus may be normal or manifest moderate venous dilatation, with later tortuosity (Fig. 3.19F); as with corkscrew conjunctival vessels, this is not pathognomonic (see Ch. 13).
- **Investigation.** CT and MRI may demonstrate prominence of the superior ophthalmic vein (Fig. 3.20A) and diffuse enlargement of extraocular muscles (Fig. 3.20B), though these may only be visible with a direct fistula. Orbital Doppler imaging may show abnormal flow patterns, particularly in the superior orbital vein. Definitive diagnosis may involve selective catheter digital subtraction angiography, especially in mild dural fistulae, though CT and MRI angiography can be useful.

Treatment

Ocular complications may require specific measures in addition to treatment of the fistula itself. Neurological subspecialist opinion should be sought at an early stage, even if features are mild, as some fistula patterns (e.g. cortical venous drainage) carry a high risk of stroke.

- **Direct.** Most carotid–cavernous fistulae are not life-threatening; the organ at major risk is the eye. Surgery is indicated if spontaneous closure does not occur. A

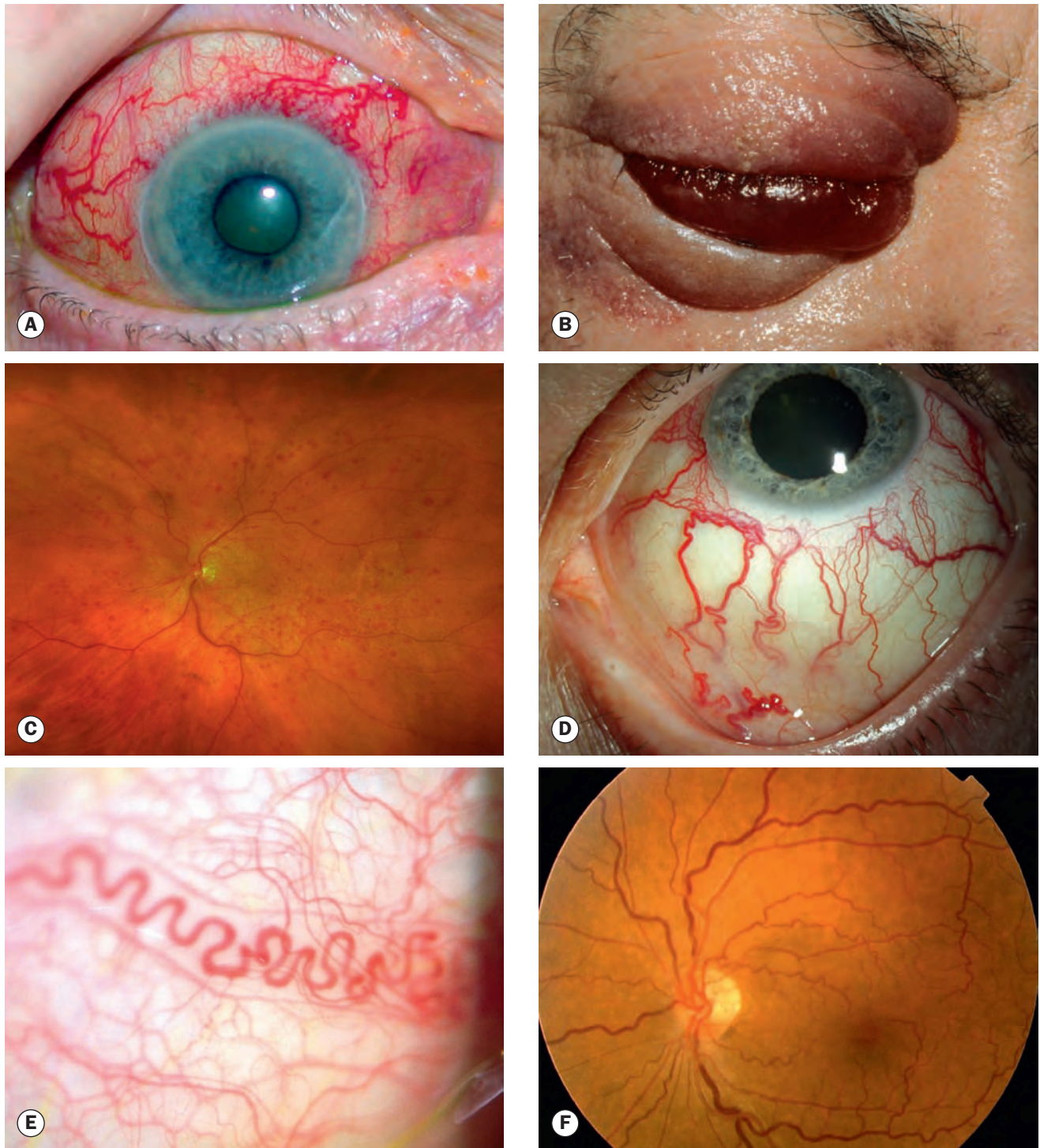


Fig. 3.19 Carotid–cavernous fistula. (A) Marked epibulbar vascular dilatation in an established direct fistula; (B) haemorrhagic chemosis in an acute direct fistula; (C) acute fundus appearance in a moderate direct fistula; (D) mild epibulbar vascular dilatation in an indirect fistula; (E) corkscrew conjunctival vessel; (F) retinal venous tortuosity

(Courtesy of S Chen – figs A and F; C Barry – figs C and D)

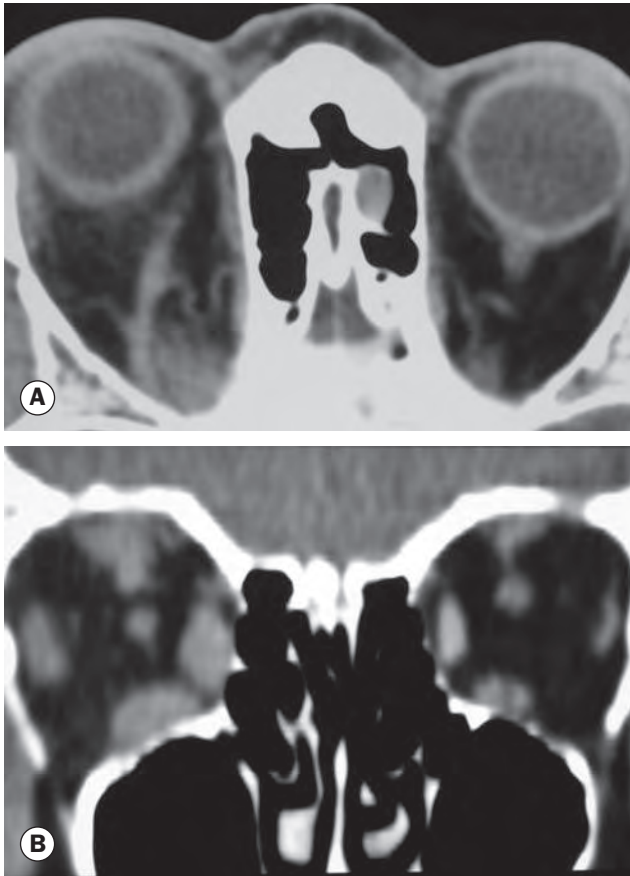


Fig. 3.20 CT in direct carotid–cavernous fistula. **(A)** Axial image shows enlargement of the right superior ophthalmic vein; **(B)** coronal view shows enlargement of extraocular muscles on the right

post-traumatic fistula is much less likely to close on its own than a spontaneous fistula because of higher blood flow. Treatment is likely to consist of a transarterial approach to repair the artery (e.g. coil – Fig. 3.21, other) or occlude the involved sinus (e.g. coil, balloon, other). Craniotomy for arterial repair is occasionally needed.

- **Indirect.** If required, treatment usually involves transvenous occlusion of the involved sinus. Spontaneous closure or occluding thrombosis sometimes (up to 50%) occurs; intermittent carotid compression under specialist supervision has been reported to increase the likelihood that this will take place.

CYSTIC LESIONS

Dacryops

A dacryops is a frequently bilateral cyst of the lacrimal gland that is thought to develop from a dilated obstructed duct. A round cystic lesion protrudes into the superior fornix from the palpebral lobe of the gland (Fig. 3.22), and may present with inflammation. The possibility of a malignant tumour should always be considered. Treatment involves excision or marsupialization, with histopathological analysis.

Dermoid cyst

Introduction

An orbital dermoid cyst is a choristoma (a mass of histologically normal tissue in an abnormal location) derived from displacement of ectoderm to a subcutaneous location along embryonic lines of closure. Dermoids are lined by keratinized stratified squamous epithelium (like skin), have a fibrous wall and contain dermal

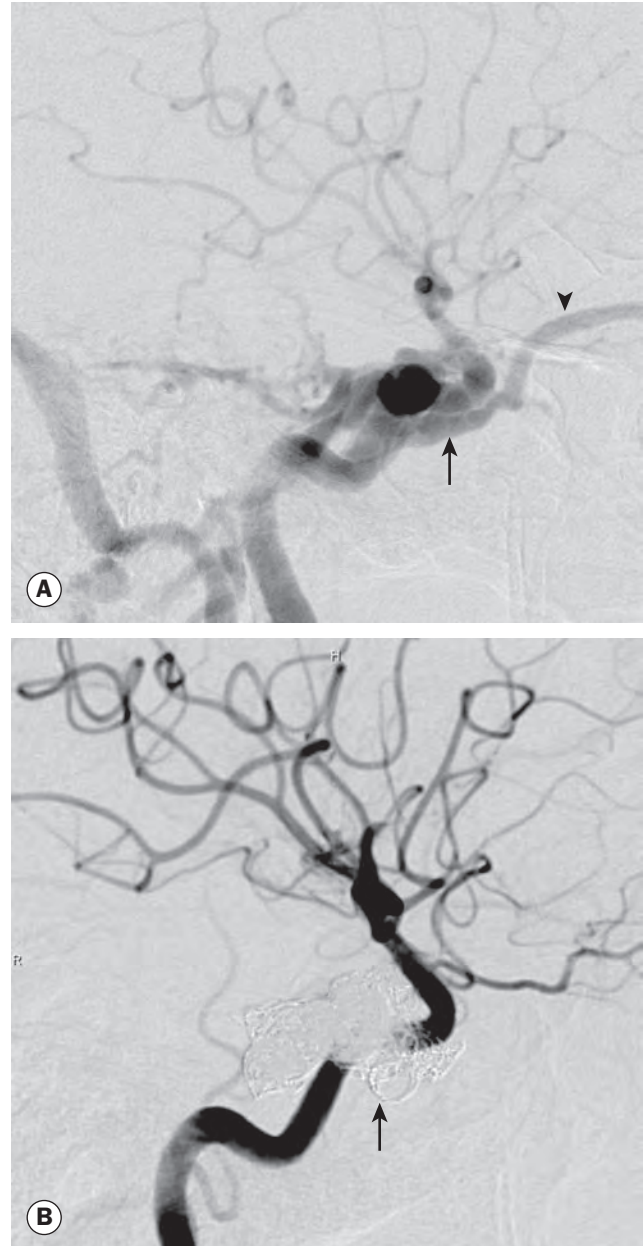


Fig. 3.21 Coil embolization of a direct carotid–cavernous fistula. **(A)** Early arterial phase catheter angiogram shows filling of the cavernous sinus (arrow) and superior ophthalmic vein (arrowhead); **(B)** following deposition of coils in the cavernous sinus – the fistula is closed and there is no retrograde flow in the superior ophthalmic vein

(Courtesy of J Trobe, from 'Neuro-ophthalmology', in Rapid Diagnosis in Ophthalmology, Mosby 2008)

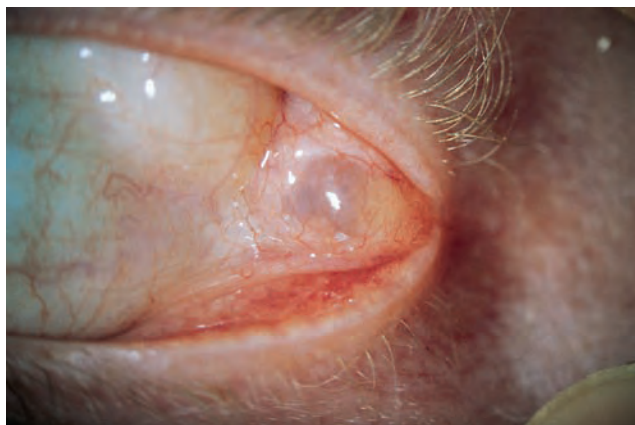


Fig. 3.22 Dacryops

appendages such as sweat glands, sebaceous glands and hair follicles; epidermoid cysts do not contain adnexal structures. Dermoids may be 'superficial' or 'deep', located anterior or posterior to the orbital septum respectively. Epibulbar dermoids and dermolipomas are related lesions (see Ch. 12).

Diagnosis

Dermoid cysts are one of the most frequently encountered orbital tumours in children.

- **Symptoms**
 - A superficial orbital dermoid cyst presents in infancy with a painless nodule, most commonly located in the superotemporal and occasionally the superonasal part of the orbit.
 - A deep dermoid cyst presents in adolescence or adult life with a gradually increasingly protruding eye, or acutely with an inflamed orbit due to rupture.
- **Signs**
 - Superficial: a firm round smooth non-tender mass 1–2 cm in diameter (Fig. 3.23A), mobile under the skin but usually tethered to the adjacent periosteum. The posterior margins are easily palpable, denoting a lack of deeper origin or extension.
 - Deep: proptosis, dystopia or a mass lesion with indistinct posterior margins (Fig. 3.23B).
- **Investigation**
 - Superficial: imaging shows a well-circumscribed heterogeneous cystic lesion (Fig. 3.24A).
 - Deep: imaging again shows a well-circumscribed lesion (Fig. 3.24B). Some deep dermoids, associated with bony defects, may extend into the inferotemporal fossa or intracranially.

Treatment

Small lesions may be observed, bearing in mind the possibility of rupture, particularly from trauma; the inflammation can be addressed with oral steroids.

- **Superficial dermoid.** Treatment is by excision *in toto* (Fig. 3.25), taking care not to rupture the lesion, since leaking of keratin into the surrounding tissue typically results in severe granulomatous inflammation.
- **Deep dermoid.** Excision *in toto* is advisable because deep dermoids enlarge and may leak into adjacent tissues inducing inflammation, often followed by fibrosis. If incompletely excised, dermoids may recur with persistent low-grade inflammation.

Sinus mucocoele

A mucocoele (US spelling – mucocoele) develops when the drainage of normal paranasal sinus secretions is obstructed due to infection, allergy, trauma, tumour or congenital narrowing. A slowly expanding cystic accumulation of mucoid secretions and epithelial debris develops and gradually erodes the bony walls of the sinus, causing symptoms by encroachment upon surrounding tissues. Orbital invasion occurs usually from a frontal or ethmoidal



Fig. 3.23 Orbital dermoid cysts. (A) Superficial cyst left eye; (B) left deep cyst causing mild dystopia

(Courtesy of A Pearson – fig. B)

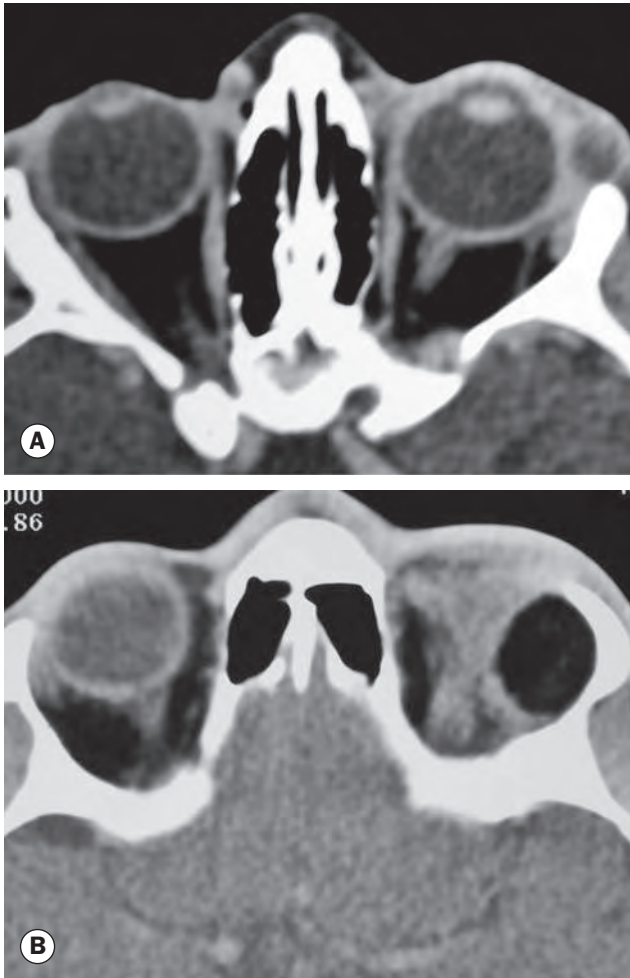


Fig. 3.24 Orbital dermoid cysts – imaging. (A) Axial CT image showing a well-circumscribed heterogeneous superficial lesion; (B) deep dermoid – CT showing a well-circumscribed cystic lesion and bone remodelling

(Courtesy of K Nischal – fig. A; A Pearson – fig. B)



Fig. 3.25 Superficial orbital dermoid cyst – appearance at surgery (orbitotomy)

(Courtesy of A Pearson)

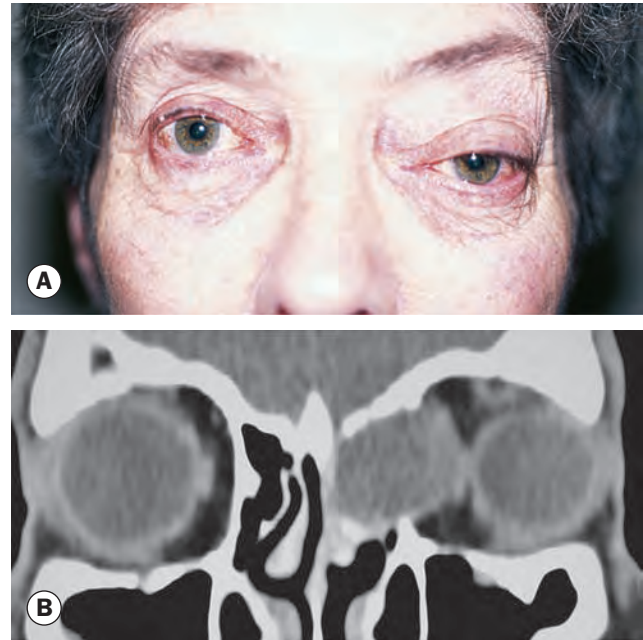


Fig. 3.26 (A) Left ethmoidal sinus mucocoele causing dystopia; (B) coronal CT shows orbital involvement and indentation of the medial rectus

mucocoele, but rarely from those arising in the maxillary sinus. Presentation is in adult life with proptosis or dystopia (Fig. 3.26A), diplopia or epiphora. Pain is uncommon unless secondary infection develops (mucopyocoele). CT shows a soft tissue mass with thinning or erosion of the bony walls of the sinus (Fig. 3.26B). Treatment involves complete excision.

Encephalocoele

An encephalocoele (US spelling – encephalocele) is formed by herniation of intracranial contents through a congenital defect of the base of the skull, and can be located at the front or back of the head. A meningocele contains only dura whilst a meningo-encephalocoele also contains brain tissue. Presentation is usually during infancy. Anterior orbital encephalocoeles involve the superomedial part of the orbit and displace the globe forwards and laterally (Fig. 3.27A), whereas posterior orbital encephalocoeles (frequently associated with neurofibromatosis type I) displace the globe forwards and downwards (Fig. 3.27B). The displacement increases on straining or crying and may be reduced by manual pressure. Pulsating proptosis may occur due to communication with the subarachnoid space but, because the communication is not vascular, there is neither a thrill nor a bruit. CT shows the bony defect responsible for the herniation (Fig. 3.27C). The differential diagnosis of anterior encephalocoele includes other causes of medial canthal swelling such as dermoid cyst and amniotocoele, and of posterior encephalocoele includes other orbital lesions that present during early life such as capillary haemangioma, juvenile xanthogranuloma, teratoma and microphthalmos with cyst.

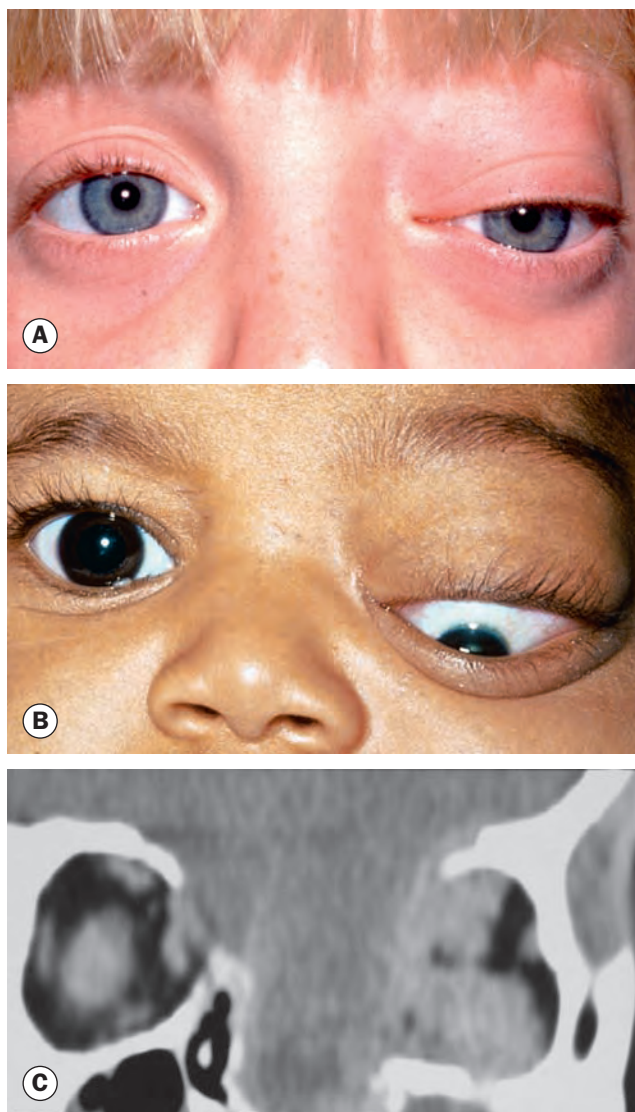


Fig. 3.27 Encephalocoele. (A) Anterior superomedial encephalocoele causing proptosis and down and out dystopia; (B) posterior encephalocoele causing proptosis and inferior dystopia; (C) coronal CT of posterior encephalocoele showing a large bony defect

(Courtesy of A Pearson – fig. C)

VASCULAR TUMOURS

Varices

Introduction

Primary orbital varices (combined venous–lymphatic malformations of the orbit – see also next topic) consist of a plexus of thin-walled distensible low-flow vein-like vessels that are commonly, though not always, intrinsic to the normal circulation. They are probably hamartomatous (hamartoma – a disorganized overgrowth of mature tissues normally present in the involved area). Associations include varices of the eyelids (Fig. 3.28A and see

Fig. 1.71) and conjunctiva (Fig. 3.28B). They commonly present at any time from early childhood to late middle age, but occasionally can be acquired later secondary to a local high-flow vascular lesion or trauma.

Diagnosis

Most cases are unilateral and the most frequent site is upper nasal. Intermittent non-pulsatile proptosis without a bruit is reported. If there is free communication with the normal circulation, reversible proptosis may be precipitated or accentuated by increasing venous pressure through coughing, straining, the Valsalva manoeuvre (Figs 3.28C and D), assuming a head-down position or external compression of the jugular veins. Imaging (e.g. MRI and magnetic resonance venography (MRV), CT, ultrasound, venography) shows a lobulated mass with variable contrast enhancement, and may demonstrate phleboliths (Fig. 3.28E) and sometimes orbital expansion (particularly in childhood) or an associated orbital wall defect. Complications include acute orbital haemorrhage, thrombosis (pain, proptosis, decreased vision) and optic nerve compression. Patients with long-standing lesions may develop atrophy of surrounding fat, giving enophthalmos with a deepened superior sulcus (Fig. 3.28F).

Treatment

Small lesions generally do not require treatment. Surgical excision is technically difficult and often incomplete because the lesions are friable and bleed easily; it can be complicated by severe orbital haemorrhage and vascular optic nerve compromise. Specialized techniques such as embolization and carbon dioxide laser surgery may be helpful adjuncts. Indications include recurrent thrombosis, pain, severe proptosis and optic nerve compression.

Lymphangioma

Introduction

Lymphangioma is a rare hamartomatous vascular tumour that tends to enlarge and infiltrate diffusely with time. Some authorities believe lymphangiomas to be a variant of venous orbital anomaly (varices) across a single spectrum, and the term combined venous–lymphatic malformations of the orbit has been suggested. Although usually isolated from the main circulation, bleeding into the lumen may occur with subsequent formation of blood-filled ‘chocolate cysts’ that may regress spontaneously with time. Presentation is usually in early childhood. Differential diagnosis is principally from orbital venous anomalies and haemangiomas. Intracranial vascular malformations can be present in association.

Diagnosis

Anterior lesions typically manifest as several soft bluish masses in the upper nasal quadrant (Fig. 3.29).

Posterior lesions may cause slowly progressive proptosis, or initially may lie dormant and later present with the sudden onset

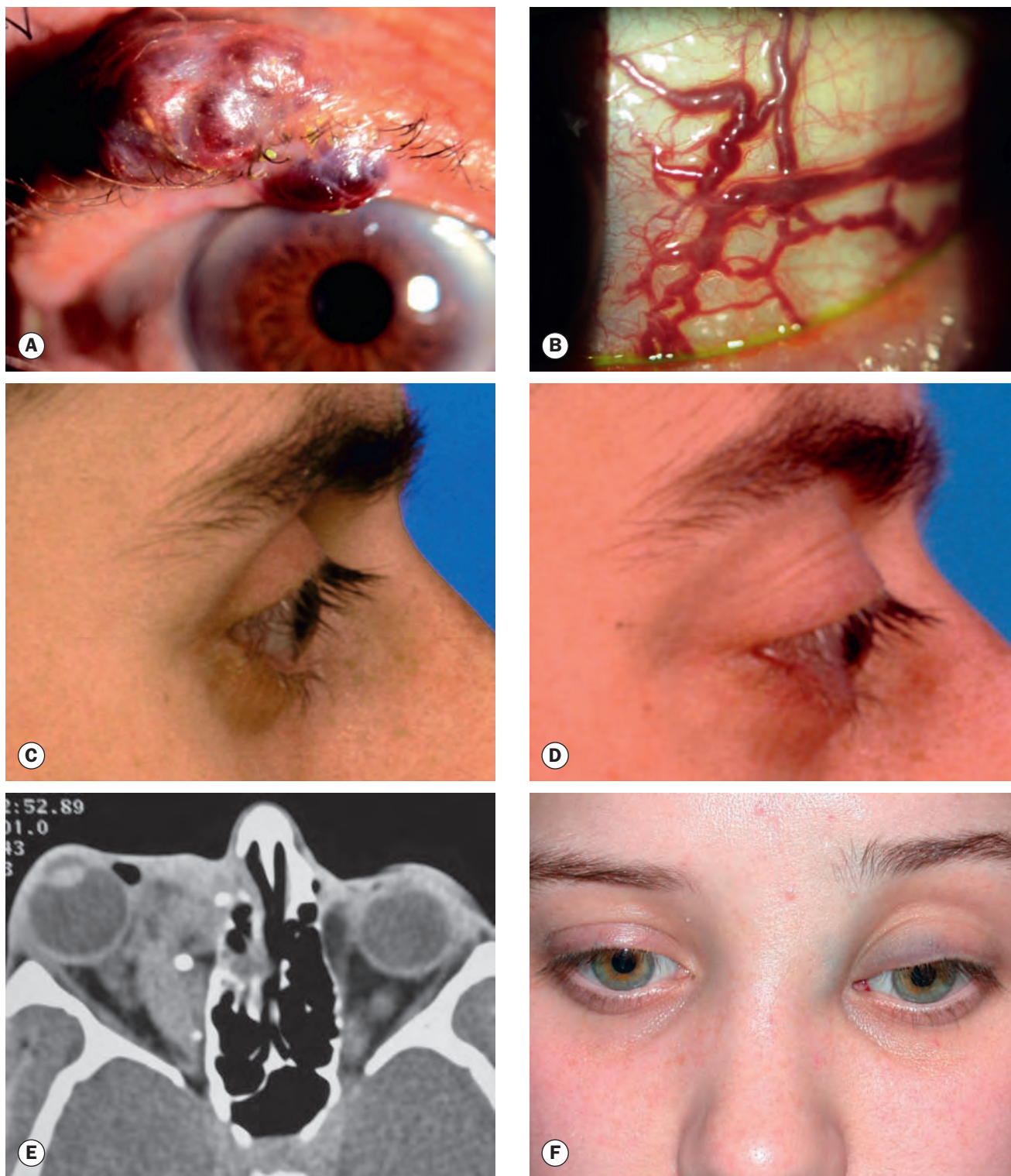


Fig. 3.28 (A) Substantial eyelid varices; (B) conjunctival varices; (C) orbital varices before Valsalva and (D) with Valsalva; (E) axial CT shows medial opacification and phleboliths; (F) left fat atrophy resulting in enophthalmos and deep superior sulcus

(Courtesy of G Rose – figs C and D; A Pearson – figs E and F)



Fig. 3.29 Anterior orbital lymphangioma with typical bluish discoloration

of painful proptosis (Figs 3.30A and B) secondary to spontaneous haemorrhage, which may be associated with optic nerve compression. Involvement of the lids, conjunctiva (Fig. 3.30C) and oropharynx (Fig. 3.30D) may be seen; intracranial lesions may also be present.

Treatment

In many cases the visual prognosis is good without treatment. Surgical excision is difficult because lesions are unencapsulated, friable, bleed easily and commonly infiltrate normal orbital tissues; repeated subtotal excision may be necessary. Persistent sight-threatening chocolate cysts can be drained or removed sub-totally by controlled vaporization using a carbon dioxide laser.

Capillary haemangioma

Introduction

Capillary haemangioma is the most common tumour of the orbit and periorbital area in childhood. Girls are affected more commonly than boys (3:1). It may present as a small isolated lesion of

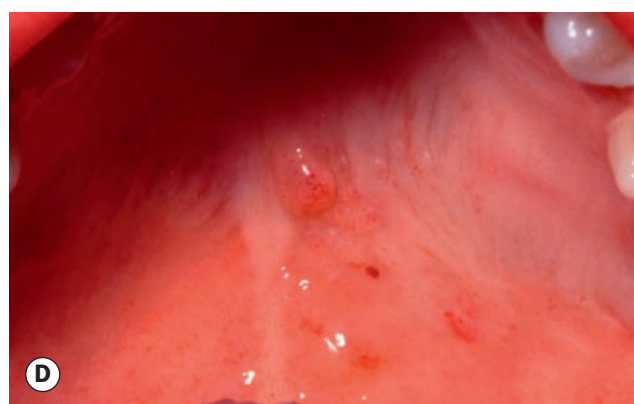
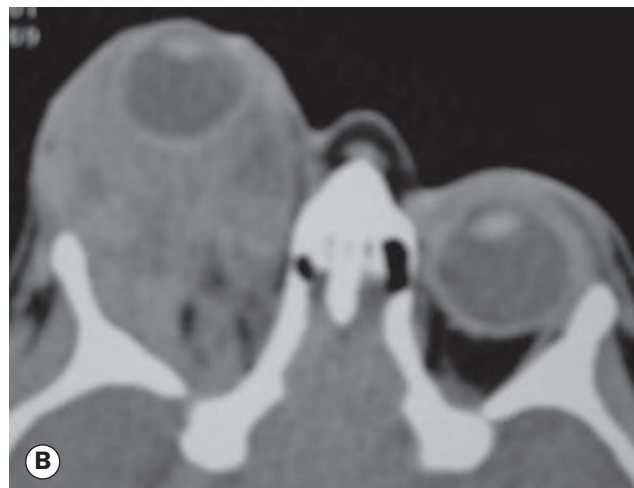


Fig. 3.30 (A) Severe proptosis due to bleeding from a posterior lymphangioma; (B) axial CT shows proptosis and orbital opacification; (C) conjunctival involvement; (D) oral lesions

(Courtesy of A Pearson – figs A and B; C Barry – fig. C)

minimal clinical significance, or as a large disfiguring mass that can cause visual impairment and systemic complications. An established tumour is composed of anastomosing small vascular channels without true encapsulation (see Fig. 1.13C); it is a hamartoma – a disorganized overgrowth of mature tissues normally present in the involved area – and believed to be due principally to endothelial cell proliferation. Large or multiple lesions may have associated visceral involvement, which can lead to serious complications such as thrombocytopenia (Kasabach–Merritt syndrome, with up to 50% mortality) and high-output cardiac failure, and systemic investigation should be considered. The incidence of infantile haemangioma in the general population is around 5%, and a small proportion of these, especially if a large facial haemangioma is present, will have PHACE (PHACES) syndrome, which includes a range of possible systemic features including eye involvement.

Diagnosis

- **Symptoms.** The lesion is usually noticed by the parent, usually in the first few months of life; approximately 30% are present at birth.
- **Signs.** Extensive underlying orbital involvement should always be ruled out in a seemingly purely superficial lesion.
 - Superficial cutaneous lesions ('strawberry naevi') are bright red (Fig. 3.31A and see Fig. 1.13A).
 - Preseptal (deeper) tumours appear dark blue or purple through the overlying skin (Fig. 3.31B and see Fig. 3.31A) and are most frequently located superiorly.
 - A large tumour may enlarge and change in colour to a deep blue during crying or straining, but both pulsation and a bruit are absent.
 - Deep orbital tumours give rise to unilateral proptosis without skin discoloration.
 - Haemangiomatous involvement of the palpebral or forniceal conjunctiva is common (Fig. 3.31C).
 - Additional haemangiomas on the eyelids (see Ch. 1) or elsewhere are common.
- **Investigation.** Imaging is generally performed for other than very small lesions, mainly to rule out more extensive orbital disease. Ultrasound shows medium internal reflectivity (Fig. 3.32A), and on MRI or CT the lesion appears as a soft tissue mass in the anterior orbit or as an extraconal mass with finger-like posterior expansions (Fig. 3.32B). The orbital cavity may show enlargement but there is no bony erosion.

Treatment

The natural course is characterized by rapid growth 3–6 months after diagnosis (Fig. 3.33), followed by a slower phase of natural resolution in which 30% of lesions resolve by the age of 3 years and about 75% by the age of 7. Treatment is indicated principally for amblyopia secondary to induced astigmatism, anisometropia, occlusion or strabismus, and less commonly for cosmesis, optic nerve compression or exposure keratopathy.

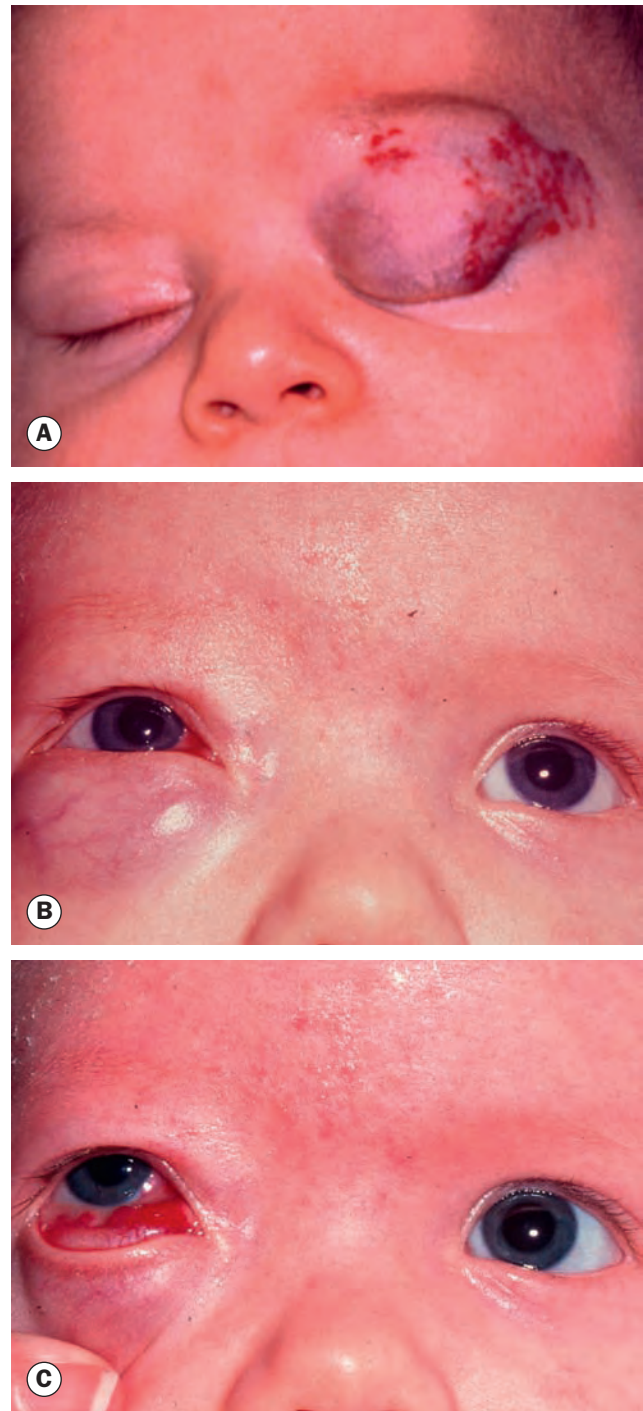


Fig. 3.31 Capillary haemangioma. (A) Large preseptal tumour causing ptosis and purple cutaneous discoloration – there is a superficial component (strawberry naevus); (B) inferior preseptal tumour; (C) involvement of forniceal conjunctiva (Courtesy of K Nischal – figs B and C)

- **Beta-blockers.** Oral propranolol is now widely used, and seems most effective in the proliferative stage; prescription and monitoring should generally be carried out by a paediatrician. Topical preparations including timolol are also under investigation, with initially favourable results.

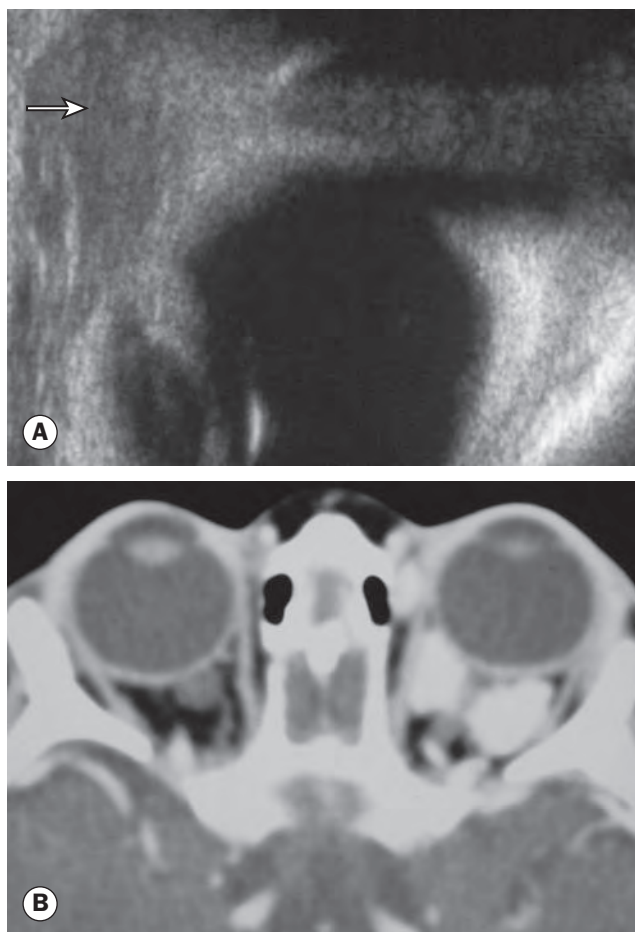


Fig. 3.32 Imaging of capillary haemangioma. (A) Ultrasound of a preseptal lesion with an intraorbital component; (B) axial enhanced CT shows a homogeneous intraconal orbital soft tissue mass

(Courtesy of K Nischal – fig. A; A Pearson – fig. B)

- **Steroids**

- Injection of triamcinolone acetonide (1–2 ml total of 40 mg/ml over several injection sites) or betamethasone (4 mg/ml) into a cutaneous or preseptal tumour is usually effective in early lesions. Regression usually begins within 2 weeks but, if necessary, second and third injections can be given after about 2 months. It is advisable not to inject deeply into the orbit for fear of causing occlusion of the central retinal artery due to retrograde introduction of the suspension. Other complications include skin depigmentation and necrosis, fat atrophy and systemic effects such as adrenal suppression.
- Topical high-potency steroids (e.g. clobetasol propionate cream) are sometimes appropriate but are slow to exert their effect.
- Systemic steroids administered daily over several weeks may be used, particularly if there is a large orbital component or a rapid onset of action is required.

- **Laser** may be used to close blood vessels in superficial skin lesions less than 2 mm in thickness.
- **Interferon alfa-2a** and **vincristine** may be used for some steroid-resistant sight-threatening lesions.
- **Local resection** with cutting cautery or carbon dioxide laser may reduce the bulk of an anterior circumscribed tumour, but is usually reserved for the late inactive stage unless a resistant tumour is sight- or life-threatening.

Cavernous haemangioma

Introduction

Cavernous haemangioma occurs in middle-aged adults, with a female preponderance of 70%; growth may be accelerated by pregnancy. It is the most common orbital tumour in adults, and is probably a vascular malformation rather than a neoplastic lesion. Although it may develop anywhere in the orbit, it most frequently occurs within the lateral part of the muscle cone just behind the globe, and behaves like a low-flow arteriovenous malformation.



Fig. 3.33 Growth of capillary haemangioma. (A) At presentation; (B) several months later

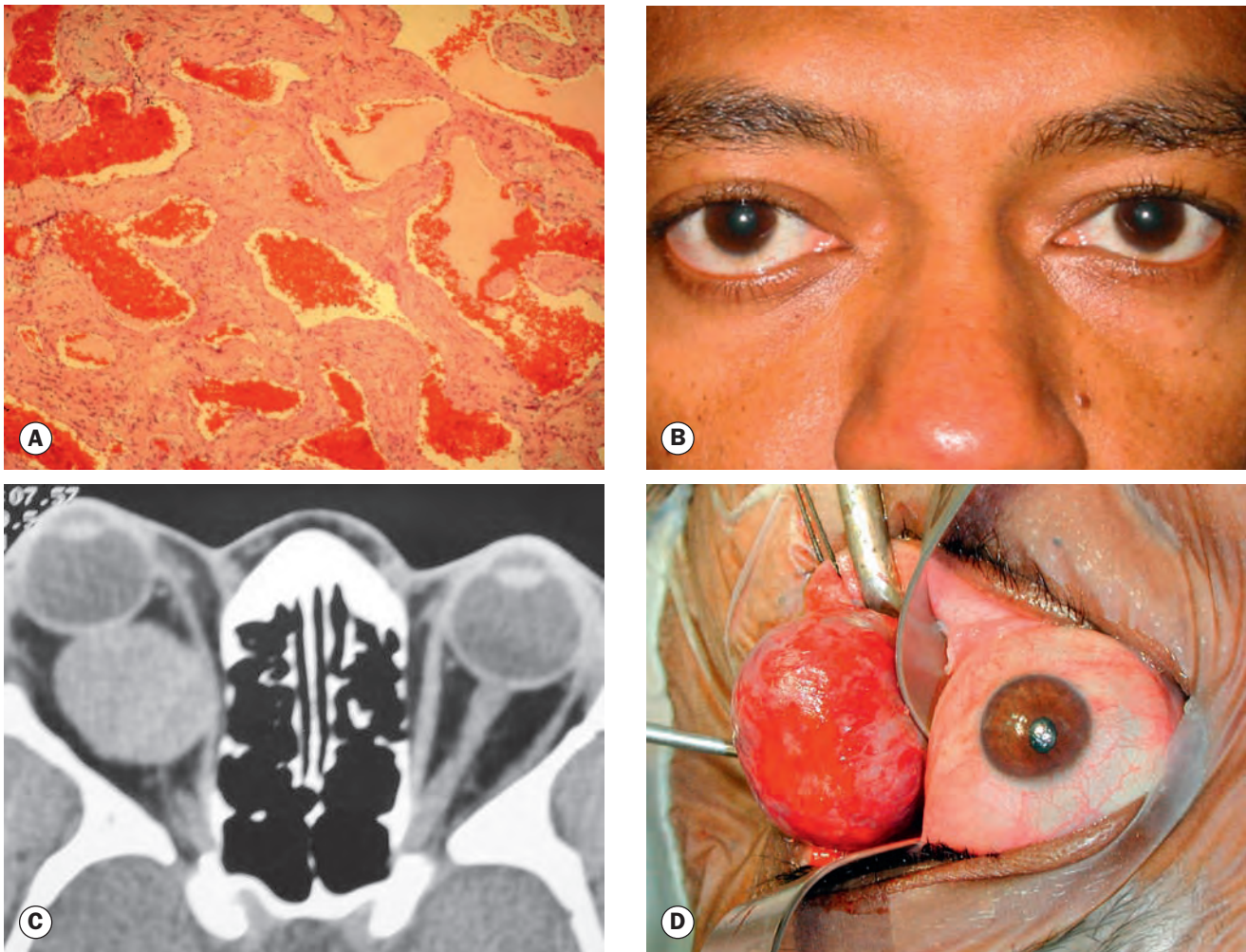


Fig. 3.34 Cavernous haemangioma. (A) Histology shows congested variably sized endothelial-lined vascular channels separated by fibrous septa; (B) right axial proptosis; (C) axial CT shows a well-circumscribed retrobulbar oval lesion and proptosis; (D) the tumour is encapsulated and relatively easy to remove

(Courtesy of A Pearson – figs B, C and D)

Histology shows endothelial-lined vascular channels of varying size separated by fibrous septa (Fig. 3.34A).

Diagnosis

- **Symptoms.** Slowly progressive unilateral proptosis; bilateral cases are very rare.
- **Signs**
 - Axial proptosis (Fig. 3.34B), which may be associated with optic disc oedema and choroidal folds.
 - A lesion at the orbital apex may compress the optic nerve without causing significant proptosis; gaze-evoked transient blurring of vision may occur.
 - There may be impairment of extraocular muscle excursion.
- **Investigation.** CT (Fig. 3.34C) and MRI show a well-circumscribed oval lesion, usually within the muscle cone. There is only slow contrast enhancement. Ultrasound is also useful.

Treatment

Many cavernous haemangiomas are detected by chance on scans performed for unrelated reasons and observation alone is often appropriate. Symptomatic lesions require surgical excision in most cases because they gradually enlarge. The cavernous haemangioma, unlike its capillary counterpart, is usually well-encapsulated and relatively easy to remove (Fig. 3.34D).

LACRIMAL GLAND TUMOURS

Pleomorphic lacrimal gland adenoma

Introduction

Pleomorphic adenoma (benign mixed-cell tumour) is the most common epithelial tumour of the lacrimal gland and is derived



Fig. 3.35 Pleomorphic lacrimal gland adenoma. **(A)** Histology shows glandular tissue and squamous differentiation with keratin formation; **(B)** inferonasal dystopia due to a tumour arising from the orbital lobe; **(C)** eyelid swelling without dystopia; **(D)** eversion of the upper eyelid reveals the tumour; **(E)** coronal CT showing an orbital lobe lesion (Courtesy of J Harry and G Misson, from *Clinical Ophthalmic Pathology*, Butterworth-Heinemann 2001 – fig. A; A Pearson – figs B and E)

from the ducts and secretory elements including myoepithelial cells. On histopathology, the inner layer of cells forms glandular tissue that may be associated with squamous differentiation and keratin production (Fig. 3.35A); the outer cells undergo metaplastic change leading to the formation of myxoid tissue. Young to middle-aged adults are the predominantly affected group.

Diagnosis

- **Symptoms.** Painless slowly progressive proptosis or swelling in the superolateral eyelid, usually of more than a year's duration. Old photographs may reveal an abnormality many years prior to presentation.
- **Signs**
 - Orbital lobe tumour presents as a smooth, firm, non-tender mass in the lacrimal gland fossa with inferonasal

dystopia (Fig. 3.35B); posterior extension may cause proptosis, ophthalmoplegia and choroidal folds.

- Palpebral lobe tumour is less common and tends to grow anteriorly causing upper lid swelling without dystopia (Fig. 3.35C); it may be visible to inspection (Fig. 3.35D).
- **Investigation.** CT shows a round or oval mass, with a smooth outline and indentation but not destruction of the lacrimal gland fossa (Fig. 3.35E). The lesion may indent the globe, and calcification may be shown.

Treatment

Treatment involves surgical excision. If the diagnosis is strongly suspected, it is wise to avoid prior biopsy to prevent tumour seeding into adjacent orbital tissue, although this may not always

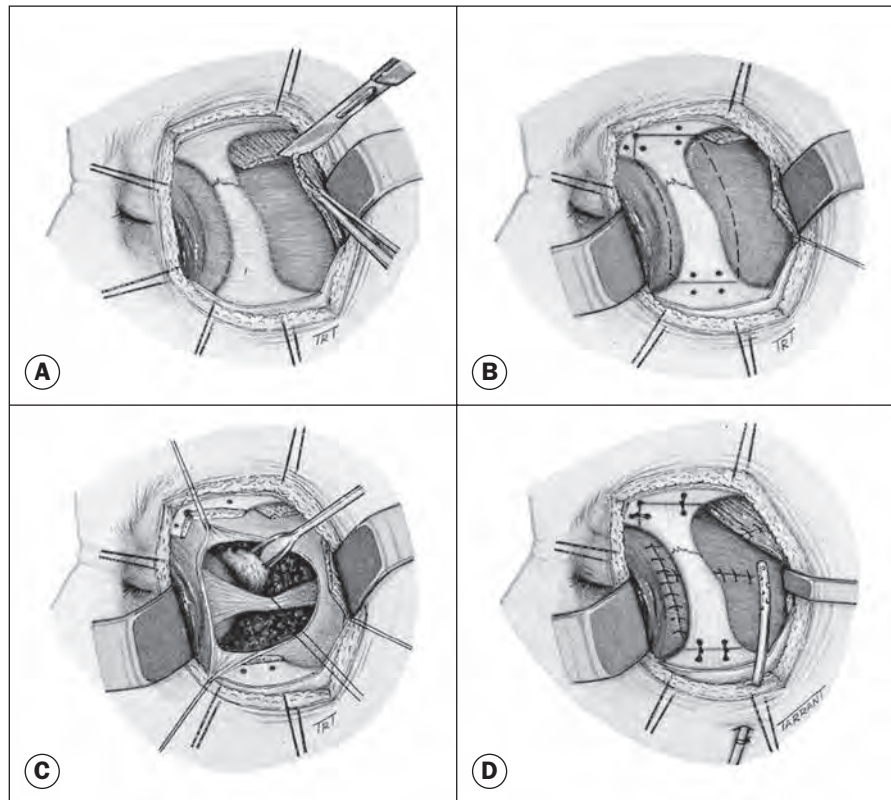


Fig. 3.36 Lateral orbitotomy. (A) Incision of temporalis muscle; (B) drilling of underlying bone for subsequent wiring; (C) removal of the lateral orbital wall and the tumour; (D) repair of the lateral orbital wall

be possible in the context of diagnostic uncertainty. Tumours of the palpebral lobe are usually resected, along with a margin of normal tissue, through an anterior (trans-septal) orbitotomy. Those of the orbital portion are excised through a lateral orbitotomy:

1. The temporalis muscle is incised (Fig. 3.36A).
2. The underlying bone is drilled for subsequent wiring (Fig. 3.36B).
3. The lateral orbital wall is removed and the tumour excised including a margin of adjacent tissue and periorbita (Fig. 3.36C).
4. The lateral orbital wall (Fig. 3.36D) and temporalis are repaired. The prognosis is excellent provided excision is complete and without disruption of the capsule. Incomplete excision or preliminary incisional biopsy may result in seeding of the tumour into adjacent tissues, with recurrence and occasionally malignant change.

Lacrimal gland carcinoma

Lacrimal gland carcinoma is a rare tumour that carries a high morbidity and mortality. In order of frequency the main histological types are adenoid cystic (50%), pleomorphic adenocarcinoma, mucoepidermoid and squamous cell. Histopathology shows nests of basaloid cells with numerous mitoses (Fig. 3.37A). The peak incidence is in middle-aged adults.

Diagnosis

- **Symptoms.** A malignant mixed-cell tumour presents in three main clinical settings:

- After incomplete or piecemeal excision of a benign pleomorphic adenoma, followed by one or more recurrences over a period of several years with eventual malignant transformation.
- As a long-standing proptosis or swollen upper lid that suddenly starts to increase.
- Without a previous history of a pleomorphic adenoma as a rapidly growing lacrimal gland mass, usually of several months' duration.
- The history is shorter than that of a benign tumour.
- Pain is a frequent feature of malignancy but may also occur with inflammatory lesions.
- **Signs**
 - A mass in the lacrimal area causing inferonasal dystopia.
 - Posterior extension, with involvement of the superior orbital fissure, may give rise to epibulbar congestion, proptosis, periorbital oedema and ophthalmoplegia (Fig. 3.37B).
 - Hypoaesthesia in the region supplied by the lacrimal nerve.
 - Optic disc swelling and choroidal folds.
- **Investigation**
 - CT shows a globular lesion with irregular serrated edges, often with contiguous erosion or invasion of bone (Fig. 3.37C). Calcification is commonly seen within the tumour.

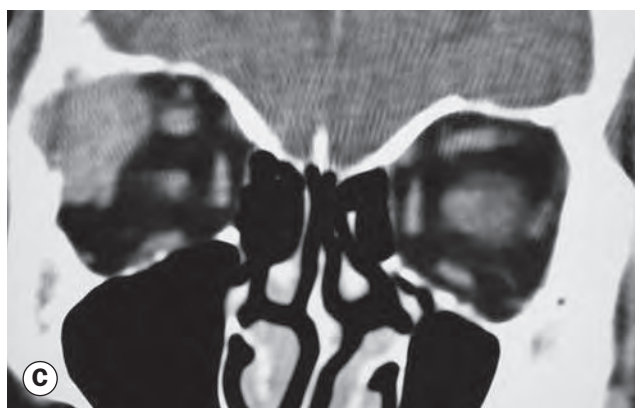
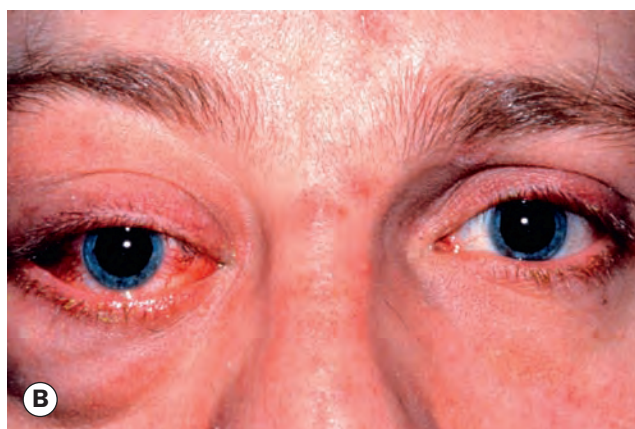
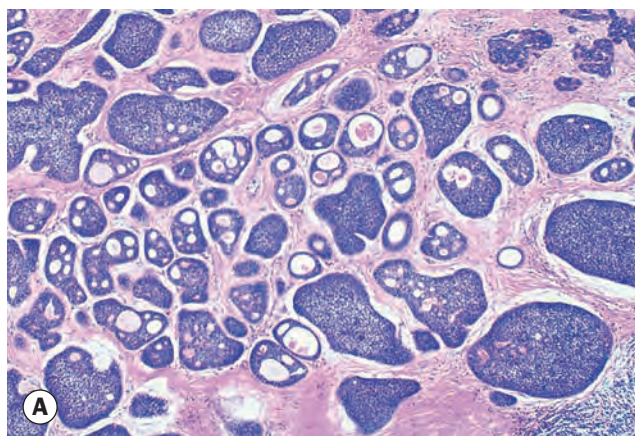


Fig. 3.37 Lacrimal gland carcinoma. **(A)** Histology of adenoid cystic carcinoma shows nests of basaloid cells with solid and cribriform areas; **(B)** dystopia, proptosis, periorbital oedema and epibulbar congestion due to extension involving the superior orbital fissure; **(C)** coronal CT shows contiguous erosion of bone and spotty calcification in the tumour

(Courtesy of J Harry and G Misson, from *Clinical Ophthalmic Pathology*, Butterworth-Heinemann 2001 – fig. A; A Pearson – fig. C)

- Biopsy is necessary to establish the histological diagnosis. Subsequent management depends on the extent of tumour invasion of adjacent structures as seen on imaging.
- Neurological assessment is mandatory because adenoid-cystic carcinoma exhibits perineural spread and may extend into the cavernous sinus.

Treatment

Treatment involves excision of the tumour and adjacent tissues. Extensive tumours may require orbital exenteration or midfacial resection, but the prognosis for life is frequently poor. Radiotherapy combined with local resection may prolong life and reduce pain. Adjuvant intra-arterial chemotherapy and/or brachytherapy may be utilized in some cases.

NEURAL TUMOURS

Optic nerve glioma

Introduction

Optic nerve glioma is a slowly growing, pilocytic astrocytoma that typically affects children (median age 6.5 years); histopathology shows spindle-shaped pilocytic (hair-like) astrocytes and glial filaments (Fig. 3.38A). The prognosis is variable; some have an indolent course with little growth, while others may extend intracranially and threaten life. Approximately 30% of patients have associated neurofibromatosis type I (NF1 – see Ch. 19) and in these patients the prognosis is generally superior. Malignant glioma (glioblastoma) is rare, has a very poor prognosis, and usually occurs in adult males.

Diagnosis

- **Symptoms**
 - Slowly progressive visual loss, followed later by proptosis, although this sequence may occasionally be reversed.
 - Acute loss of vision due to haemorrhage into the tumour can occur, but is uncommon.
- **Signs**
 - Proptosis is often non-axial, with temporal or inferior dystopia (Fig. 3.38B).
 - The optic nerve head, initially swollen, subsequently becomes atrophic.
 - Opticociliary collaterals (see Fig. 3.6C) and other fundus signs such as central retinal vein occlusion are occasionally seen.
 - Intracranial spread to the chiasm and hypothalamus may develop.
- **Investigation**
 - MRI effectively demonstrates the tumour, and may show intracranial extension if present.
 - CT in patients with associated NF1 shows a fusiform enlargement of the optic nerve with a clear-cut margin produced by the intact dural sheath (Fig. 3.38C). In patients without NF1 the nerve is more irregular and shows low-density areas.

Treatment

As the tumour is intrinsic to the optic nerve, resection means that all vision will be lost in the operated eye.

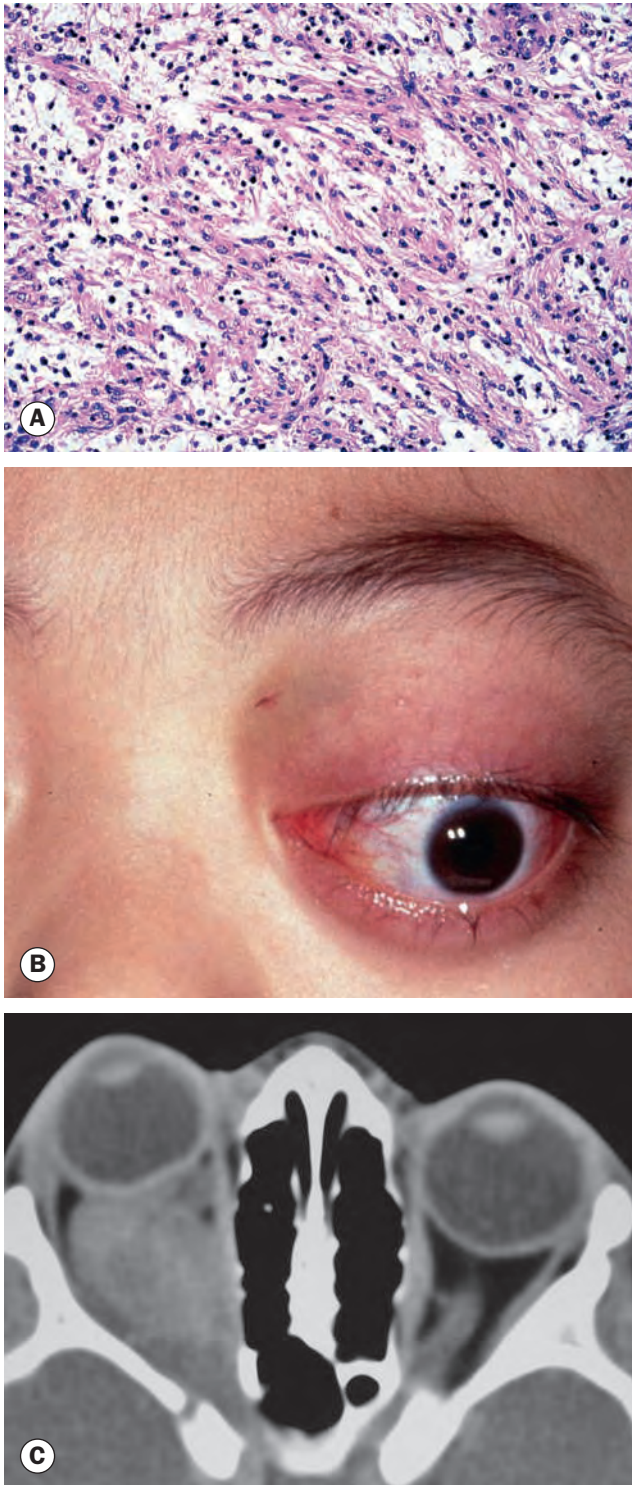


Fig. 3.38 Optic nerve glioma. **(A)** Histopathology – spindle-shaped pilocytic astrocytes and glial filaments; **(B)** proptosis with inferior dystopia; **(C)** axial CT showing fusiform optic nerve enlargement

(Courtesy of J Harry – fig. A; K Nischal – fig. B; A Pearson – fig. C)

- **Observation** may be considered in patients with a typical pilocytic astrocytoma on imaging in whom the tumour is confined to the orbit, especially if there is good vision and no significant cosmetic impairment; serial MRI is important if this option is chosen. Spontaneous regression has been reported, usually in NF1, but is very rare.
- **Surgical excision** with preservation of the globe is indicated in those with large or growing tumours where complete resection of the tumour can be achieved, particularly if vision is poor and proptosis significant. A key goal is to prevent chiasmal involvement, and an intracranial approach may be necessary to achieve adequate resection.
- **Radiotherapy** may be combined with chemotherapy for tumours with extension that precludes complete surgical excision.

Optic nerve sheath meningioma

Introduction

Optic nerve sheath meningioma is a benign tumour arising from meningotheial cells of the arachnoid villi surrounding the intraorbital, or less commonly the intracanalicular, portion of the optic nerve. In some cases the tumour merely encircles the optic nerve whilst in others it invades the nerve, growing along the fibrovascular pial septa. However, about two-thirds of all meningiomas affecting the optic nerve arise from extension of primarily intracranial lesions. Primary optic nerve sheath meningiomas are less common than optic nerve gliomas and, as with other meningiomas, typically affect middle-aged women. Histopathologically, meningotheial (irregular lobules of meningotheial cells separated by fibrovascular strands – Fig. 3.39A) and psammomatous (psammoma bodies among proliferating meningotheial cells – Fig. 3.39B) types are distinguished. The prognosis for life is good in adults, although the tumour may be more aggressive in children, in whom 25% occur. They are more common in neurofibromatosis type II (NF2).

Diagnosis

- **Symptoms** typically consist of gradual visual impairment in one eye. Transient obscurations of vision may occur.
- **Signs.** The classic (Hoyt–Spencer) triad consists of progressive visual loss, optic atrophy and opticociliary shunt vessels, although the simultaneous occurrence of all three signs in one individual is actually uncommon. The usual sequence of involvement is the opposite of that seen in tumours that develop outside the dural sheath:
 1. Optic nerve dysfunction and chronic disc swelling followed by atrophy.
 2. Opticociliary collaterals (30%); these regress as optic atrophy supervenes.
 3. Restrictive motility defects, particularly in upgaze (Fig. 3.39C).
 4. Proptosis.

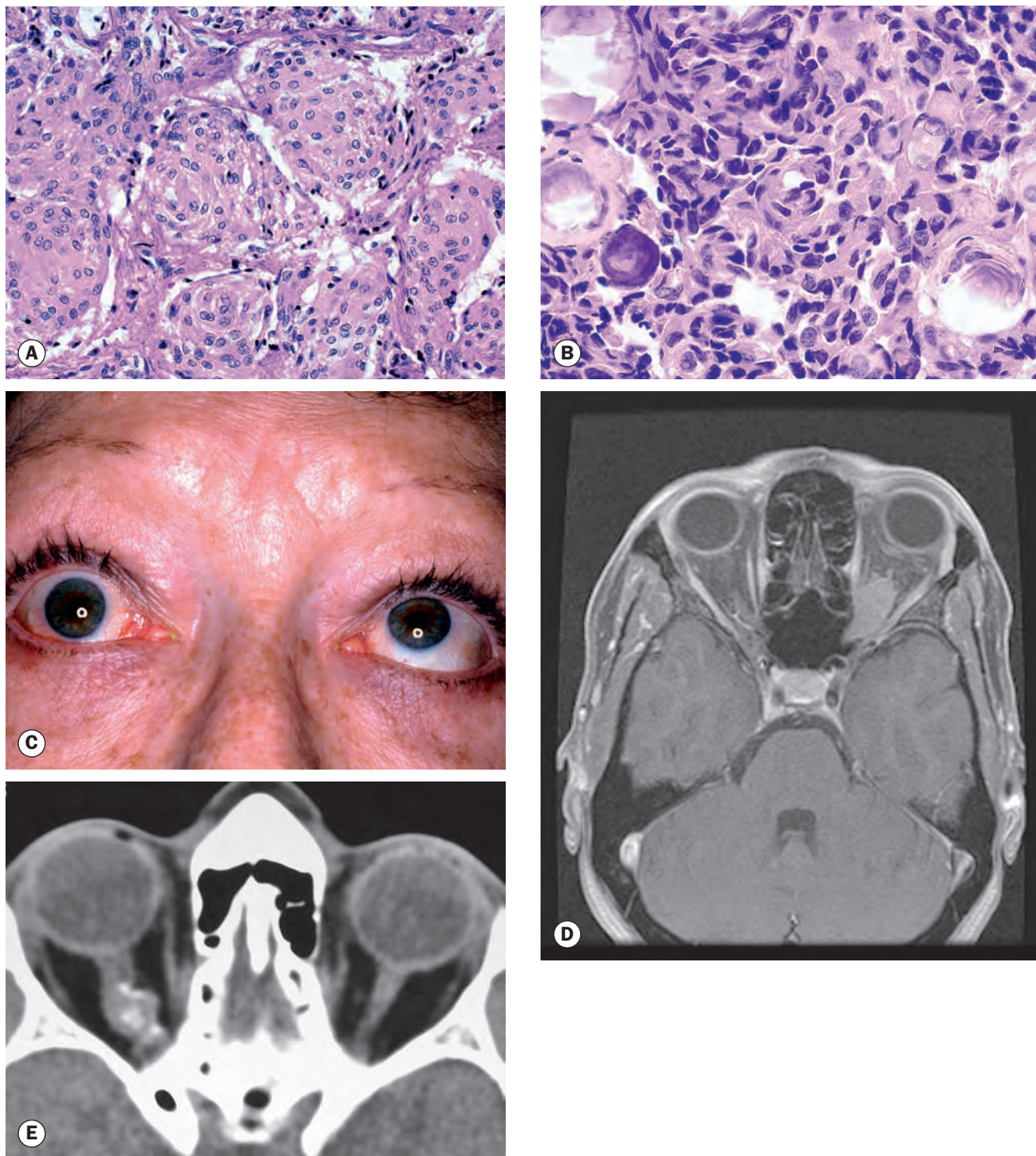


Fig. 3.39 Optic nerve meningioma. (A) Meningoethelial histopathology; (B) psammomatous histopathology; (C) defective elevation of the right eye; (D) MRI showing lesion associated with left optic nerve; (E) axial CT of a small tumour showing calcification

(Courtesy of J Harry and G Misson, from *Clinical Ophthalmic Pathology*, Butterworth-Heinemann 2001 – figs A and B; A Pearson – fig. E)

• Investigation

- MRI is the investigation of choice (Fig. 3.39D).
- CT shows thickening and calcification of the optic nerve (Fig. 3.39E).
- Ultrasonography (especially coronal) may be useful.

Treatment

Treatment may not be indicated in a middle-aged patient with a slowly growing lesion, but excision is required for an aggressive tumour, particularly if the eye is blind or there is a risk of

intracranial extension. Attempts at optic nerve-sparing surgery commonly fail but may be considered on a case basis. Fractionated stereotactic radiotherapy may be appropriate as a vision-sparing approach, or as adjunctive treatment following surgery.

Plexiform neurofibroma

Plexiform neurofibroma is the most common peripheral neural tumour of the orbit. It occurs almost exclusively in association with NF1. Presentation is in early childhood with periorbital swelling; classically, involvement of the eyelids causes mechanical ptosis with a characteristic S-shaped deformity (see Fig. 1.16), but diffuse involvement of the orbit with disfiguring hypertrophy of periocular tissues may occur. On palpation the involved tissues are said to resemble a 'bag of worms'. Malignant change can occur, and should be suspected if there is rapid change; radiotherapy may promote this. Treatment is often unsatisfactory and complete surgical removal is extremely difficult. Orbital surgery should be avoided when possible because of the intricate relationship between the tumour and important structures.

Isolated neurofibroma

Isolated (localized) neurofibroma is less common than plexiform neurofibroma; about 10% of patients have NF1. Presentation is in the third or fourth decades with insidious mildly painful proptosis, usually not associated with visual impairment or ocular motility dysfunction. Excision is commonly straightforward because the tumour is well-circumscribed and relatively avascular.

LYMPHOMA

Introduction

Lymphomas of the ocular adnexa constitute approximately 8% of all extranodal lymphomas. The majority of orbital lymphomas are non-Hodgkin, and most of these (80%) are of B-cell origin. Those affected are typically older individuals. The condition may be primary, involving one or both orbits only, or secondary if there are one or more identical foci elsewhere in the body; a substantial proportion of apparently primary lesions will develop disease elsewhere within a few years. The course is variable and relatively unpredictable. In some patients histological features raise suspicion of malignancy and yet the lesion resolves spontaneously or with steroid treatment. Conversely, what appears to be reactive lymphoid hyperplasia may be followed by the development of lymphoma. Small lesions and those involving only the conjunctiva have the best prognosis. Conjunctival and intraocular lymphomas are discussed in Chapter 12.

Diagnosis

The onset is characteristically insidious.

- **Symptoms.** An absence of symptoms is common, but may include discomfort, double vision, a bulging eye or a visible mass.

- **Signs**

- Any part of the orbit may be affected (Fig. 3.40A); anterior lesions may be palpated, and generally have a rubbery consistency (Fig. 3.40B).
- Occasionally the lymphoma may be confined to the conjunctiva or lacrimal glands, sparing the orbit.
- Local lymph nodes should be palpated, but systemic evaluation by an appropriate specialist is required.

- **Investigation**

- Orbital imaging, usually with MRI (Fig. 3.40C).
- Biopsy is usually performed to establish the diagnosis.
- Systemic investigation to establish the extent of disease.

Treatment

Radiotherapy is used for localized lesions, and chemotherapy for disseminated disease and some subtypes. Immunotherapy (e.g. rituximab) is a newer modality that may assume a dominant role in the future. Occasionally a well-defined orbital lesion may be resected.

RHABDOMYOSARCOMA

Introduction

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma of childhood: 40% develop in the head and neck, and it is the most common primary orbital malignancy in children but is still a rare condition; 90% occur in children under 16 and the average age of onset is 7 years. The tumour is derived from undifferentiated mesenchymal cells that have the potential to differentiate into striated muscle. Various genetic predispositions have been identified, including variants of the *RB1* gene responsible for retinoblastoma. Four subtypes are recognized:

- **Embryonal** constitutes the majority (85%) of orbital lesions. Cells may show light microscopic features of striated muscle differentiation. Embryonal usually carries a good prognosis.
- **Alveolar** makes up most of the balance of orbital RMS. Fewer cells show skeletal muscle differentiation than embryonal, and the prognosis is worse. Particular chromosomal translocations are characteristic on cytogenetic analysis of biopsy material.
- **Botyroid** (4%) and **pleomorphic** RMS are much less common in the orbit.

Diagnosis

- **Symptoms.** Rapidly progressive unilateral proptosis is usual, and may mimic an inflammatory condition such as orbital cellulitis.
- **Signs**
 - The tumour is most commonly superonasal or superior, but may arise anywhere in the orbit, including inferiorly (Figs 3.41A and B). It can also arise in other tissues, such as conjunctiva and uvea.

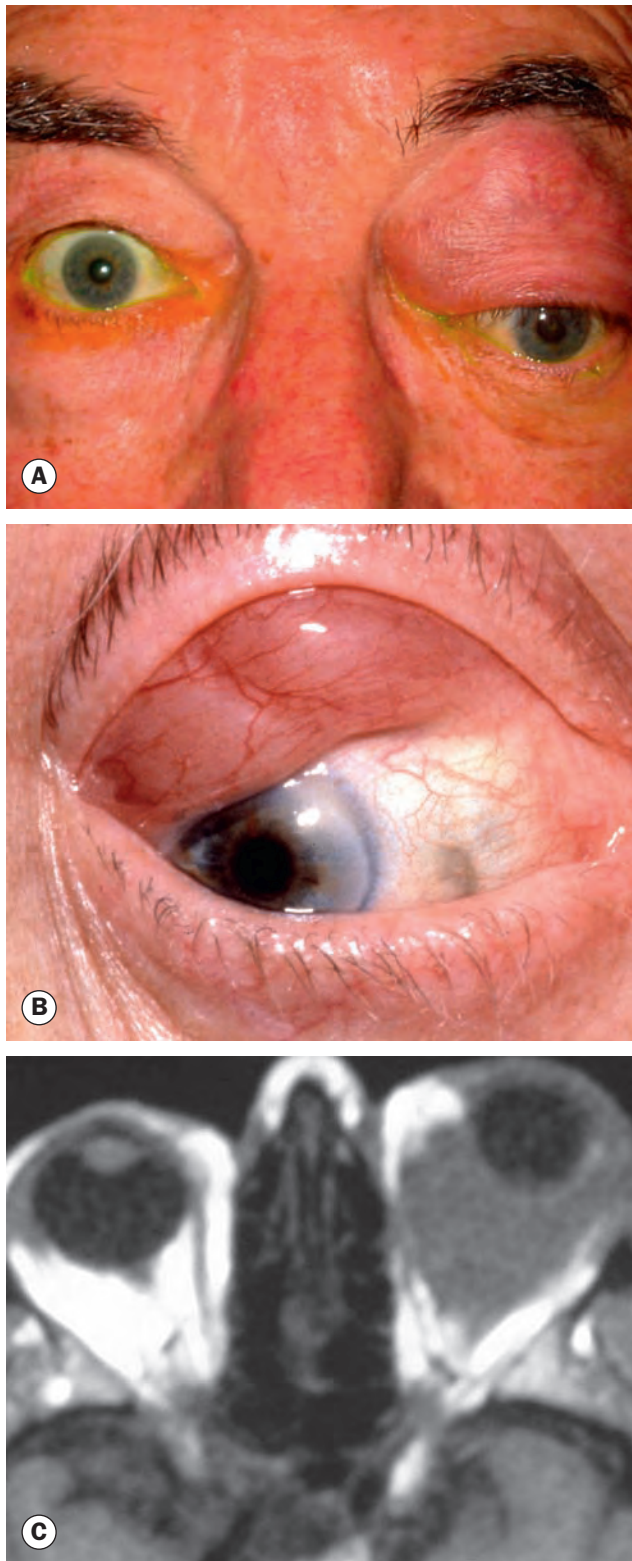


Fig. 3.40 Orbital lymphoma. (A) Involvement of the superior orbit causing proptosis and inferior dystopia; (B) anterior lesion; (C) axial T1-weighted MR of the patient in (A) shows a large orbital soft tissue mass and proptosis

(Courtesy of A Pearson – figs A and C)

- Swelling and redness of overlying skin develop but the skin is not warm (see Fig. 3.41A).
- Diplopia is frequent, but pain is less common.
- **Investigation**
 - MRI shows a poorly defined mass (Fig. 3.41C).
 - CT shows a poorly defined mass of homogeneous density, often with adjacent bony destruction (Fig. 3.41D).
 - Incisional biopsy is performed to confirm the diagnosis and establish the histopathological subtype and cytogenetic characteristics.
 - Systemic investigation for metastasis should be performed; the most common sites are lung and bone.

Treatment

Commonly used guidelines for staging and a corresponding treatment protocol were produced by the Intergroup Rhabdomyosarcoma Study Group (IRSG); treatment encompasses a combination of radiotherapy, chemotherapy and sometimes surgical debulking. The prognosis for patients with disease confined to the orbit is good.

METASTATIC TUMOURS

Adult metastatic tumours

Introduction

Orbital metastases are an infrequent cause of proptosis, and are much less common than metastases to the choroid. If the orbit is the site of initial manifestation of the tumour, the ophthalmologist may be the first specialist to see the patient. In approximate order of frequency the most common primary sites are breast (up to 70%), bronchus, prostate, skin (melanoma), gastrointestinal tract and kidney.

Diagnosis

- **Signs.** Associated with the range of tumours that can spread to the orbit, presentation can take a variety of forms.
 - Dystopia and proptosis (Fig. 3.42A) are the most common features.
 - Infiltration of orbital tissues characterized by ptosis, diplopia, brawny indurated periorbital skin and a firm orbit, with resistance to manual retropulsion of the globe.
 - Enophthalmos with scirrhous tumours.
 - Chronic inflammation.
 - Primarily with cranial nerve involvement (II, III, IV, V, VI) and only mild proptosis with orbital apex lesions.
- **Investigation**
 - Imaging: CT (Fig. 3.42B) and MRI typically show a non-encapsulated mass.
 - Fine needle biopsy is useful for histological confirmation. If this fails, open biopsy may be required.
 - A search for a primary must be carried out if the patient was not previously known to have cancer.

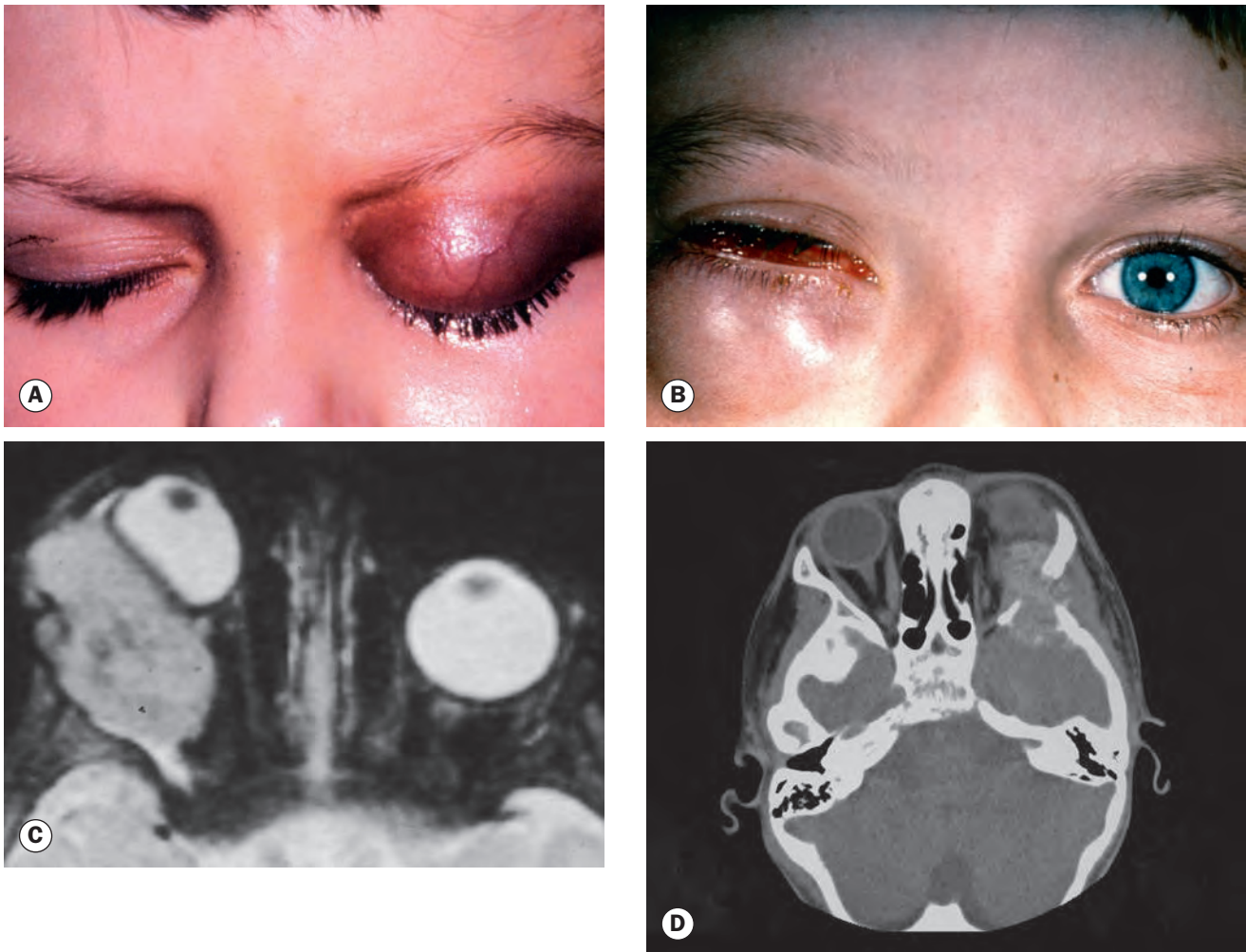


Fig. 3.41 Rhabdomyosarcoma. (A) Superiorly and (B) anteroinferiorly located lesions; (C) axial T2-weighted MR showing a large retrobulbar mass with indentation of the globe and proptosis; (D) CT showing bony destruction and intracranial spread (Courtesy of M Szreter – fig. A; A Pearson – fig. B; S Chen – fig. D)

Treatment

Treatment is aimed at preserving vision and relieving pain, because most patients die within a year (average 4 months). Radiotherapy is the mainstay of local treatment, but systemic therapy may also be of benefit. Surgical excision of the focus is occasionally carried out. Orbital exenteration is usually only performed if other modalities fail to control intolerable symptoms.

Childhood metastatic tumours

Neuroblastoma

Neuroblastoma is one of the most common childhood malignancies. It arises from neural crest-derived tissue of the sympathetic nervous system, most commonly in the abdomen (Fig. 3.43A). Presentation is usually in early childhood; in almost half of all cases the tumour is disseminated at diagnosis, when it carries a very poor prognosis. Orbital metastases may be bilateral and

typically present with the abrupt onset of proptosis accompanied by a superior orbital mass and lid ecchymosis (Fig. 3.43B).

Myeloid sarcoma

Myeloid sarcoma (granulocytic sarcoma) is a solid tumour composed of malignant cells of myeloid origin. Because the tumour may exhibit a characteristic green colour it was formerly referred to as chloroma. Myeloid sarcoma may occur as a manifestation of established myeloid leukaemia, or it may precede the disease. Orbital involvement usually presents at about age 7 years with the rapid onset of proptosis, sometimes bilateral, and can be associated with ecchymosis and lid oedema. When orbital involvement precedes systemic leukaemia, the diagnosis may be difficult.

Langerhans cell histiocytosis

Langerhans cell histiocytosis is a rare group of disorders due to clonal proliferations of histiocytes. Presentation ranges from localized disease, usually with bone destruction (eosinophilic granuloma), through multifocal bone involvement to a fulminant



Fig. 3.42 Metastatic renal carcinoma. (A) Proptosis; (B) axial CT showing a non-encapsulated retrobulbar mass
(Courtesy of A Pearson – fig. B)

systemic disease. Soft tissues are less commonly involved, but cutaneous and visceral lesions may occur. Orbital involvement consists of unilateral or bilateral osteolytic lesions and soft tissue involvement, typically in the superotemporal quadrant (Fig. 3.44). Patients with solitary lesions tend to have a more benign course, and respond well to treatment with local curettage and intralesional steroid injection or radiotherapy. Systemic disease has a worse prognosis.

Orbital invasion from adjacent structures

Sinus tumours

Malignant tumours of the paranasal sinuses, although rare, may invade the orbit and carry a poor prognosis unless diagnosed early. It is therefore important to be aware of both their otorhinolaryngological and ophthalmic features.

- **Maxillary carcinoma** is by far the most common sinus tumour to invade the orbit (Fig. 3.45).
 - Otorhinolaryngological manifestations include facial pain and swelling, epistaxis and nasal discharge.
 - Ophthalmic features include upward dystopia, diplopia and epiphora.

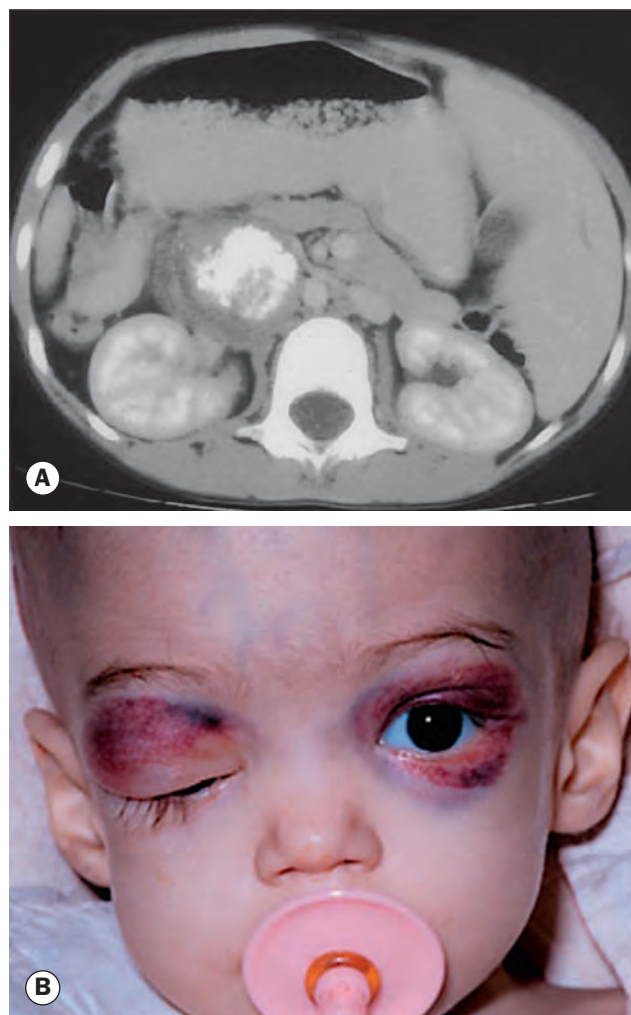


Fig. 3.43 Neuroblastoma. (A) Axial CT shows a tumour adjacent to the kidney; (B) bilateral orbital metastases
(Courtesy of B Zitelli and H Davis, from Atlas of Pediatric Physical Diagnosis, Mosby 2002)



Fig 3.44 Langerhans cell histiocytosis
(Courtesy of D Taylor)



Fig. 3.45 Advanced maxillary carcinoma showing facial swelling and upward dystopia

- **Ethmoidal carcinoma** may cause lateral dystopia.
- **Nasopharyngeal carcinoma** may spread to the orbit through the inferior orbital fissure; proptosis is a late finding.

Bony invasion

- **Intracranial meningioma** arising from the sphenoid ridge may invade the orbit by direct spread and cause proptosis (see Fig. 19.54). Occasionally tumours arising from the tuberculum sellae or olfactory groove may invade the orbit through the superior orbital fissure or optic canal.
- **Fibrous dysplasia** is a disorder in which fibrous tissue develops instead of normal bone, leading to weakening and a mass effect (Fig. 3.46A), usually in childhood or early adulthood. Within the orbital region this may cause facial asymmetry, proptosis, dystopia (Fig. 3.46B) and visual loss. Most orbital disease is due to the monostotic form; polyostotic disease is associated with endocrine disorders and cutaneous pigmentation (McCune–Albright syndrome – 10% of cases).

Orbital invasion from eyelid, conjunctival and intraocular tumours

- Orbital invasion may occur from eyelid malignancies such as basal cell carcinoma, squamous cell carcinoma or sebaceous gland carcinoma, from conjunctival tumours (e.g. melanoma – Fig. 3.47A), and from intraocular tumours such as choroidal melanoma or retinoblastoma (Fig. 3.47B).

THE ANOPHTHALMIC SOCKET

Surgical procedures

Removal of an eye or the contents of the orbit may be indicated for intraocular or orbital malignancy or where the eye is blind and

painful or unsightly. A number of different surgical and rehabilitation techniques are available.

Enucleation

Enucleation (removal of the globe) is indicated in the following circumstances:

- **Primary intraocular malignancies** where other treatment modalities are not appropriate. The tumour is left intact within the eye for histopathological examination.
- **After severe trauma** where the risk of sympathetic ophthalmitis may outweigh any prospect of visual recovery; a rare indication (see Ch. 21).
- **Blind painful or unsightly eyes** can also be managed by enucleation, although evisceration is generally considered the procedure of choice.

Evisceration

Evisceration refers to removal of the entire contents of the globe, whilst the sclera and extraocular muscles remain intact. Generally the cornea is removed (Fig. 3.48) to provide access to the ocular contents. Retention of the sclera and lack of disruption of the

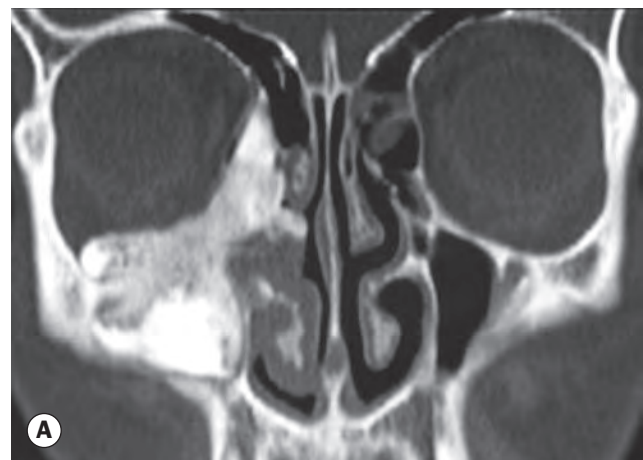


Fig. 3.46 Fibrous dysplasia of the orbit. (A) Coronal CT scan showing involvement of the floor and medial wall of the right orbit; (B) upward dystopia of the right eye

(Courtesy of A Pearson)



Fig. 3.47 (A) Orbital invasion by conjunctival melanoma; (B) orbital invasion by retinoblastoma

extraocular muscles is considered to provide somewhat better motility than is achieved after enucleation. Evisceration provides disrupted and incomplete material for histology and should not be undertaken in the presence of suspected intraocular malignancy.



Fig. 3.48 Appearance following evisceration
(Courtesy of S Chen)

Exenteration

Exenteration involves removal of the globe together with the soft tissues of the orbit (Figs 3.49A and B). Indications include:

- **Orbital malignancy**, either primary or where a tumour has invaded the orbit from the eyelids, conjunctiva, globe or adnexa, when other forms of treatment have a very poor chance of success. Anteriorly sited tumours may allow relative sparing of posterior orbital tissue, and posterior tumours may allow sparing of eyelid skin to line the socket (Fig. 3.49C). Following exenteration, prostheses can be attached to the surrounding skin with adhesive, mounted on glasses (Fig. 3.50), or secured with osseointegrated magnets mounted on the orbital rim bones. The socket may be lined with skin or a split-skin graft, or left to heal by secondary intention.
- **Non-malignant disease** such as orbital mucormycosis is a rare indication.

Rehabilitation

Cosmetic shell

A cosmetic shell is an ocular prosthesis that is used to cover a phthisical or unsightly eye. The shell can restore volume, and often provides a good cosmetic appearance, with reasonable motility as a result of transmitted movements from the globe.

Orbital implants

Enucleation or evisceration leads to a reduction in volume of the orbital contents. A large prosthetic eye without an underlying orbital implant does not provide a satisfactory solution, due to stretching of the lower lid under its weight and to poor motility. An implant is usually inserted during the surgery at which the eye is removed, though secondary placement can be performed later or a previously inserted implant exchanged. Implant materials may be solid ('non-integrated', e.g. silicone, acrylic) or porous ('integrated', e.g. polyethylene, hydroxyapatite). Fibrovascular ingrowth into the latter facilitates motility of an overlying prosthesis; a peg can also be inserted into porous implants to improve later motility, though the peg must be covered *in situ* by socket tissue and cannot attach directly to the overlying prosthesis. The motility of unpegged implants is also usually good, particularly if donor sclera or a mesh wrap is used and the extraocular muscles secured to the surface.

- **Post-enucleation socket syndrome (PESS)** is caused by failure to correct the volume deficit adequately. It is characterized by a deep upper lid sulcus, ptosis, enophthalmos (Fig. 3.51) and backwards rotation of the top of the prosthesis.
- **Extrusion** (Fig. 3.52) is a significant concern with all implants. Careful placement of an implant, ensuring it is sufficiently deep and is well covered with vascularized tissue, is more important than the choice of implant material.



Fig. 3.49 Exenteration. (A) Including eyelid removal, 2 days postoperatively; (B) patient in (A) 6 months later; (C) with sparing of the eyelids
(Courtesy of S Chen – figs A and B; A Pearson – fig. C)



Fig. 3.50 (A) Healed exenteration; (B) prosthesis attached to glasses
(Courtesy of A Pearson)



Fig. 3.51 Right post-enucleation socket syndrome (PESS)



Fig. 3.52 Extruding orbital implant

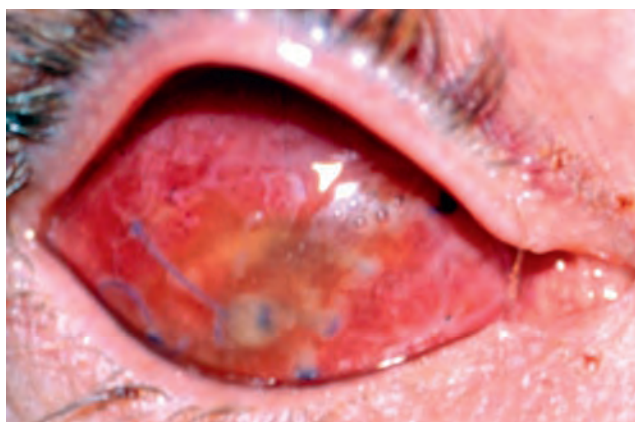


Fig. 3.53 Conformer in place



Fig. 3.54 A selection of artificial eyes
(Courtesy of C Barry)

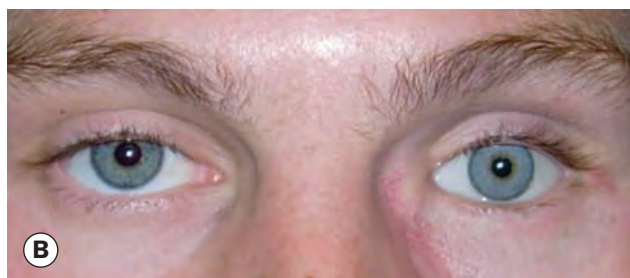


Fig. 3.55 Prosthetic eye. (A) Empty socket; (B) matching prosthesis in place
(Courtesy of S Chen)



Fig. 3.56 Crouzon syndrome. (A) Proptosis, midfacial hypoplasia and mandibular prognathism; (B) proptosis and hypertelorism – a 'V' exotropia is also shown

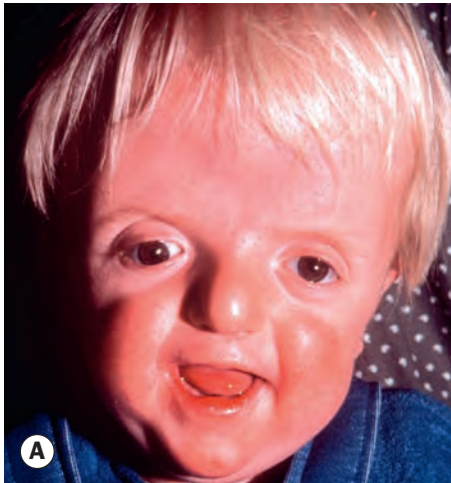


Fig. 3.57 Apert syndrome. **(A)** Mild shallow orbits, midfacial hypoplasia and 'parrot-beak' nose; **(B)** syndactyly

Ocular prosthesis

After enucleation or evisceration a conformer (Fig. 3.53) made of silicone or acrylic material is placed to support the conjunctival fornices and remains in place until the socket is fitted with an

artificial eye (Fig. 3.54). Initial socket impression moulds can usually be taken at around 6–8 weeks postoperatively and a temporary artificial eye placed whilst waiting for manufacture of a prosthesis shaped to fit the individual socket and matched to the fellow eye (Fig. 3.55).

CRANIOSYNOSTOSES

The craniosynostoses are a group of rare congenital conditions in which an abnormally shaped skull results from premature closure of skull sutures.

- **Crouzon syndrome** features a short anteroposterior skull diameter, with midfacial hypoplasia giving a prominent lower jaw (Fig. 3.56A); proptosis due to shallow orbits and hypertelorism (wide orbital separation) are the most conspicuous ocular features (Fig. 3.56B). Vision-threatening complications include exposure keratopathy and optic atrophy, mechanisms including chronic papilloedema and cerebral hypoperfusion secondary to sleep apnoea. Strabismus ('V' exotropia – see Fig. 3.56B), ametropia and amblyopia can occur, and other ocular associations have been reported. Inheritance is usually autosomal dominant (AD); allelic variants in the gene *FGFR2* are responsible.
- **Apert syndrome** is the most severe of the craniosynostoses. Oxycephaly (conical skull), midfacial hypoplasia with a beaked nose and low-set ears (Fig. 3.57A), syndactyly (Fig. 3.57B) and developmental delay (30%) may be present. Shallow orbits, proptosis and hypertelorism are generally less pronounced than in Crouzon syndrome, but the same vision-threatening complications occur. Inheritance can be AD, but in the majority of cases the condition is sporadic and associated with older parental age. As with Crouzon syndrome, it is frequently the result of mutations in *FGFR2*.
- **Pfeiffer syndrome** features midfacial hypoplasia and down-slanting palpebral fissures. Ocular features are similar to Apert syndrome. Inheritance is AD with genetic heterogeneity.